

SLIDE 1 - TITLE: Arthritis Mutilans in psoriatic arthritis

CAPTION: This patient is a 49 year old Hispanic female with a history of arthralgia for 5 years that presented for further evaluation. She was found to have synovitis of the MCPs and PIPs. Inflammatory markers were elevated, but CCP and RF were negative. X-rays of the hands revealed: on the right, erosive changes at the third through fifth DIPs, the third and fifth PIP and the third through fifth CMCs. The erosions have a pencil in cup appearance. There is volar subluxation at the fifth DIP. There is an erosion at the trapezium as well. On the left hand: there were erosions at the third through fifth PIPs, the fourth and fifth PIPs and 4th and 5th CMCs some of which also have a pencil in cup configuration. There is dorsal subluxation of the third DIP. In the setting of negative RF/CCP, and the deformities described above the patient was diagnosed with psoriatic arthritis.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: X-ray of the hands

SLIDE 2 - TITLE: Fulminant Eosinophilic myocarditis in a dermatomyositis patient.

CAPTION: A 53 years old female with a muscle biopsy proven diagnosis of dermatomyositis for 25 years presented to the emergency department complaining of acute onset of shortness of breath, chest discomfort, malaise and vomiting. She described the discomfort as mid-sternal, dull, radiating to the back, not aggravated by exercise or food intake. On the physical examination, she seemed to be in mild discomfort, had a heliotrope rash and decrease in proximal muscle strength on both upper and lower extremities. In less than 48 hours, she had a 2-fold increase in troponin (22 at admission to 58 on day 2), and her systolic cardiac function deteriorated from 45% to 10%, with diffuse hypokinesis on transthoracic echocardiogram. At that point she received a biventricular assisted device to support her cardiac function. Endomyocardial biopsy was consistent with necrotizing eosinophilic myocarditis, a thorough work up ruled out an infectious etiology. She was immunosuppressed with steroids and rituximab, received IV immunoglobulins, and required pressors to maintain her cardiac function. This hospital course was complicated by ischemic bowel with intra-abdominal bleeding that required multiple surgical interventions and a cerebral vascular accident. After 10 weeks of hospital course, in the setting of multiple organ failure, the family decided to withdraw care. Cardiac involvement is rare in dermatomyositis, but an important cause of death. The role of immunosuppressive therapy remains controversial. The correlation of overall severity, disease activity and cardiac manifestations is also controversial. Physicians should maintain a high index of suspicion in patients with dermatomyositis.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: Cardiac MRI: There is diffuse nonhomogeneous increased T2 signal throughout the myocardium of the left ventricle as well as along the pericardium. Post contrast evaluation: There is diffuse areas of patchy enhancement along the left ventricle and portions of the right ventricle. Involvement of the at the end pericardium is noted. Patchy areas of delayed enhancement of the myocardium as well as enhancement of the pericardium, compatible with severe myo-pericarditis. Severe global biventricular hypokinesis with an estimated LVEF of 20%

HISTOPATHOLOGY DESCRIPTION: H&E-stained sections of myocardium show intense inflammation of right ventricular myocardium with extension into adipose tissue (Panel A). Under high magnification (Panel B), numerous eosinophils can be seen within the infiltrate. Confluent destruction of cardiac myocytes is also evident. (Original magnification: A 100x; B 400x)

SLIDE 3 - TITLE: Temporomandibular joint synovial osteochondromatosis

CAPTION: Axial CT (left) and axial T2-weighted MRI (right) show numerous osteochondral bodies within a distended right temporomandibular joint.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: CT without contrast and T2 MRI

SLIDE 4 - TITLE: Cutaneous Polyarteritis Nodosa(PAN): Bilateral leg ulcers

CAPTION: 51 y/o male with history of cutaneous polyarteritis nodosa (PAN), presented with extensive deep ulcers in both legs and the dorsum of the right foot. There are exposures of the tendons on the lateral side of the right leg.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: None

SLIDE 5 - TITLE: Diffuse Cutaneous Systemic Sclerosis(dcSSc): "Salt- and - pepper" appearance of the chest and neck

CAPTION: 52 y/o female with Scl-70(topoisomerase I) positive, diffuse cutaneous systemic sclerosis (dcSSc) presenting with diffuse hypopigmentation(vitiligo-like) and hyperpigmentation of the skin ("Salt- and- pepper" appearance) in the chest and neck.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: None

SLIDE 6 - TITLE: Diffuse Cutaneous Systemic Sclerosis (dcSSc): "Salt- and- pepper appearance" of the skin

CAPTION: 52 y/o female with Scl-70(topoisomerase I) positive, diffuse cutaneous systemic sclerosis (dcSSc) presenting with diffuse hypopigmentation(vitiligo-like) and hyperpigmentation of the skin ("Salt- and- pepper" appearance) in the abdomen.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: None

SLIDE 7 - TITLE: Tumoral calcinosis in systemic sclerosis

CAPTION: A 60-year-old woman presented with a 3-year history of pain and swelling of the both fingers. She had a history of Raynaud's phenomenon. Antinuclear antibody (discrete speckled)and anti-centromere antibody were positive. Hand X-ray showed an acroosteolysis of the digital phalanges. Femur X-ray showed extensive tumoral calcinosis on the periarticular region of the right hip. The patient was diagnosed with systemic sclerosis.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: Thigh Rt MRI (Gd enhanced) Multilobulating cloud like calcifications are seen at right gluteus minimus, tensor fascia lata with displacement of gluteus medius, rectus femoris and iliopsoas muscles. It surrounded right hip joint. Pherepheral enhancement is seen around the calcified nodules.

SLIDE 8 - TITLE: Large-vessel giant cell arteritis with secondary polymyalgia rheumatica

CAPTION: 70 year-old female presented low-grade fever, neck and pelvic girdle pain, jaw claudication and pulseless. Laboratory examinations showed that C-reactive protein was 11 mg/dl and ESR was 123 mm/hr. FDG-PET/CT findings demonstrated bilateral subclavian and carotid arteritis and aortitis in addition to bursitis and enthesitis of spinous process and pelvic girdle. We diagnosed as large-vessel giant cell arteritis (GCA) with secondary polymyalgia rheumatica (PMR). Glucocorticoid therapy was started, and not only these symptoms and but also abnormal findings of FDG-PET/CT were improved.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: FDG-PET/CT showed that bilateral subclavian and carotid arteritis and aortitis (as Giant cell arteritis) in addition to bursitis and enthesitis of spinous process and pelvic girdle as polymyalgia rheumatica (PMR).

SLIDE 9 - TITLE: Erythema nodosum and Oral ulcer in Behcet's syndrome

CAPTION: A 40 year old Iraqi woman with recurrent oral and genital ulcers was evaluated for skin lesions on her legs that were tender, consistent with erythema nodosum. She also complained of joint pains. Her symptoms did not respond to a combination of colchicine and azathioprine and so, she was started on infliximab with good repsonse.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: None

SLIDE 10 - TITLE: Linear Hypopigmentation and Skin Atrophy Following Peri-Enthesal Steroid Injection for Lateral Epicondylitis

CAPTION: A 33-year-old woman received a single peri-enthesal injection of 40 mg triamcinolone acetonide for left lateral epicondylitis. Tenderness of lateral epicondyle and pain with resisted wrist extension and full pronation resolved following the therapeutic steroid injection. Two months after the injection, she noted a 0.5 cm hypopigmented spot at the site of the injection (not pictured). Four months following the injection, the hypopigmented macule had increased in size to 1.5 cm with a streaky halo effect as well as a hypopigmented projection extending proximally in a linear fashion (attached photo). She refused skin biopsy.

ADDITIONAL CLINICAL INFORMATION: Linear cutaneous hypopigmentation and atrophy following intra-articular and intralesional steroid injections were thought to be the results of lymphatic uptake of the corticosteroid crystals (1). Etiology of hypopigmentation with corticosteroids relates to decreased melanocyte function (2). References: 1. Kikuchi I, Horikawa S. Letter: perilymphatic atrophy of the skin. Arch Dermatol. 1975;111:795-6. 2. Venkatesan P, Fangman WL. Linear hypopigmentation and cutaneous atrophy following intra-articular steroid injections for de Quervain's tendonitis. J Drugs Dermatol. 2009;8:492-3.

X-RAY DESCRIPTION: None

SLIDE 11 - TITLE: Rheumatoid Arthritis (mutilans)

CAPTION: A woman with rheumatoid arthritis (mutilans type). Note severe bone defect around shoulders and elbows.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: X-ray

SLIDE 12 - TITLE: Takayasu's Aortitis

CAPTION: 17 y.o. Asian Female who complained of pain when eating. Symptoms were present for more than one year with a great deal of family strife as the patients parents thought she was 'depressed, willful' since previous doctors failed to make a diagnosis. Examination of the neck revealed a loud bruit and a CT angiogram of the abdomen showed massive involvement of virtually all of the vessels. The patient was started on cyclophosphamide with significant improvement in her laboratory values but not in her symptoms. She developed severe abdominal pain and was taken to emergency surgery where near total small bowel necrosis was found. She died later that day.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: CT angiogram

SLIDE 13 - TITLE: Gout and Pseudogout present in knee aspirate

CAPTION: Polarizing light microscope picture of a monosodium urate (MSU) crystal and a calcium pyrophosphate dehydrate (CPPD) crystal in the same field. Arrow shows the direction of the polarization and points away from the MSU crystal and points toward the CPPD crystal. The MSU crystal is needle shaped and negatively birefringent (yellow) parallel to the polarizer. The CPPD crystal is rhomboid shaped and positively birefringent (blue) parallel to the polarizer.

ADDITIONAL CLINICAL INFORMATION: Aspirate from a red, hot, swollen knee joint.

X-RAY DESCRIPTION: None

SLIDE 14 - TITLE: Calcific Periarthritis in a Young Female Athlete

CAPTION: A 27 year old healthy female presented with left great toe pain clinically mimicking gout. Imaging revealed calcific periarthritis.

ADDITIONAL CLINICAL INFORMATION: The patient was a previously healthy 27 year old female participating in daily exercise consisting of biking and running on alternating days. She initially developed minor discomfort in the left great toe with weight bearing which prompted her to stop running, however, she continued biking. After a particularly long 25 mile bike her toe pain escalated and the left great toe became red, warm, and swollen for 2

weeks. Her physical exam was consistent with podagra however she was premenopausal making gout unlikely. X-ray imaging of the foot showed calcific peri-arthritis. The patient did not respond to Naproxen therapy and thus was given a short prednisone taper with resolution of her symptoms.

X-RAY DESCRIPTION: X-Ray; Calcification plantar to hallux IP joint suggests hydroxyapatite crystal deposition possibly related to flexor hallucis tendon. No findings of inflammatory or erosive arthropathy

SLIDE 15 - TITLE: Myelodysplastic syndrome: Bone marrow findings of iron overload

CAPTION: A 58 year old female with myelodysplastic syndrome. Dual-sequence MR imaging of the pelvis. Bone marrow is very low signal intensity in both T1 and T2 fat-saturated images due to reticuloendothelial accumulation of iron secondary to multiple blood transfusions leading to hemosiderosis (iron overload). The excess iron causes severe shortening of the T2 relaxation time resulting in conspicuous lowering of marrow signal intensity on all pulse sequences. Similar findings occur in other organs subject to iron overload such as the liver.

ADDITIONAL CLINICAL INFORMATION: A 58-year-old woman presented with shortness of breath and right thigh pain. Patient had a longstanding history of anemia and was recently diagnosed with Myelodysplastic syndrome (MDS). She required multiple transfusions, 19 U of packed RBCs in three and a half years, along with iron and aranesp therapy. Patient also had a history of stage III non-small-cell lung cancer, status post chemotherapy and chemoradiation, chronic renal disease, hypothyroidism and hypertension. Rheumatology was consulted to evaluate myositis in the right thigh. MRI of the pelvis revealed incidental low signal intensity bone marrow secondary to hemosiderosis. Myositis and cellulitis responded well to antibiotic therapy.

X-RAY DESCRIPTION: MRI of the Pelvis with T1-weighted and T2-weighted, fat-saturated images 1. Soft tissue edema of the buttocks bilaterally and myositis of the anterior thigh muscles are incompletely shown. The patient's myelodysplastic syndrome lead to iron accumulation in the hematopoietic marrow with hemosiderosis causing severe T2 relaxation time shortening and resultant low signal intensity of affected bone marrow on all magnetic resonance images. 2. No septic arthritis or osteomyelitis; imaging findings consistent with cellulitis of the buttocks and myositis of the right thigh anterior compartment.

SLIDE 16 - TITLE: Dactylitis as the first manifestation of psoriatic arthritis in a 2 year old child

CAPTION: 2 year old girl with swelling of the left fifth finger for five months and dactylitis on MRI, diagnosed with JIA of psoriatic arthritis subtype.

ADDITIONAL CLINICAL INFORMATION: A 2-year-1-month-old girl of Turkish origin developed a 'sausage-like' swelling in her left fifth finger accompanied by mild pain, as well as swelling and onycholysis in several toes. Contrast MRI imaging of the hand demonstrates enhancement around the metacarpophalangeal, proximal and distal interphalangeal joint of the fifth finger accompanied by tenosynovitis of the flexor tendon of that finger. This involvement of the distal and proximal interphalangeal joints, together with tendon sheath involvement, is a typical finding of dactylitis, allowing the diagnosis of juvenile psoriatic arthritis according to the ILAR criteria for juvenile idiopathic arthritis, underscored by psoriasis in a second degree relative. This patient substantially improved on treatment with naproxen and methotrexate and continues to do well on last follow-up.

X-RAY DESCRIPTION: T1 weighted MRI with contrast. Contrast MRI imaging of the hand demonstrates enhancement around the metacarpophalangeal, proximal and distal interphalangeal joint of the fifth finger accompanied by tenosynovitis of the flexor tendon of that finger.

SLIDE 17 - TITLE: Bisphosphonate induced femoral fracture

CAPTION: 58 year old female with seropositive RA and osteoporosis, sustained bilateral atypical femoral fractures after bisphosphonate use (for 4 years).

ADDITIONAL CLINICAL INFORMATION: Patient was on Alendronate and presented with acute onset pain and limping on right leg.

X-RAY DESCRIPTION: X -ray of bilateral femurs: There is cortical beaking and a lucent fracture line at the lateral aspect of the subtrochanteric right and left femur, consistent with a bisphosphonated-associated stress fracture.

SLIDE 20 - TITLE: Ankylosing Spondylitis; Spine Radiographs

CAPTION: 62 year old male with acute C6-C7 fracture and complete fusion of anterior and posterior spinal elements secondary to Ankylosing Spondylitis.

ADDITIONAL CLINICAL INFORMATION: 62 year old male with history of Ankylosing Spondylitis for 25 years, on NSAIDS as needed, was admitted after a mechanical fall. Patient presented with severe neck pain and numbness of both hands. He was found to have an acute fracture through C6-C7 hypertroph extending into the disc with associated spinal cord edema.

X-RAY DESCRIPTION: PA, AP and lateral views of cervical, thoracic and lumbosacral spine demonstrate complete ankylosis of anterior and posterior spinal elements, including supraspinous and interspinous ligaments with ankylosis of bilateral Sacroiliac Joints. There is also a displaced fracture through C6-C7 intervertebral disc space and C6 vertebral body with disruption of bridging ventral syndesmotic changes and C7 spinous process.

SLIDE 22 - TITLE: Libman Sacks Endocarditis

CAPTION: 27 year old female with history of SLE presented with chest pain and shortness of breath with Mitral Regurgitation (MR) secondary to Libman Sacks Endocarditis.

ADDITIONAL CLINICAL INFORMATION: Patient had a history of SLE and presented with acute onset of chest pain and shortness of breath. Echocardiogram showed verrucous vegetations on both mitral valve leaflets with severe MR and reduced LV systolic function. Endomyocardial biopsy revealed occlusive thrombus in intramural coronary vessel with no evidence of myocarditis. Patient also developed antiphospholipid syndrome with thrombocytopenia and was treated with pulse steroids, anticoagulation, plasmapheresis and cyclophosphamide.

X-RAY DESCRIPTION: None

HISTOPATHOLOGY DESCRIPTION: Transesophageal Echocardiogram showing verrucous vegetations at the tips of both mitral valve leaflets. Endomyocardial biopsy reveals an occlusive thrombus in intramural coronary vessel with no evidence of myocarditis or inflammatory cellular infiltrate (H & E stain).

SLIDE 23 - TITLE: Popliteal Synovial Cyst

CAPTION: This image shows a large fluctuant swelling along the medial aspect of the right popliteal fossa consistent with a large popliteal synovial cyst with extension into the calf. This 69 year-old man with a 7-year history of rheumatoid arthritis presented with difficulty kneeling due to tightness of right knee with flexion. Pain occurred only with squatting. There was no history of trauma. Knee X-ray showed evidence of osteoarthritis. Popliteal cysts, also known as Baker's cysts, can be seen in individuals with rheumatoid arthritis, as well as those with knee osteoarthritis. They can also be seen in individuals with meniscal tears, septic arthritis, and pigmented villonodular synovitis. In patients with rheumatoid arthritis, a popliteal cyst with dissection into the calf is a more common cause of unilateral calf swelling, as demonstrated by this case, than a deep venous thrombosis.

ADDITIONAL CLINICAL INFORMATION: This 69 year-old man with a 7-year history of seropositive, anti-CCP positive rheumatoid arthritis presented with difficulty kneeling due to tightness of right knee with flexion. Knee X-ray showed mild joint space narrowing in both the medial and lateral compartments with subchondral sclerosis consistent with mild osteoarthritis. There were no erosions. On examination, there was a large effusion in the suprapatellar pouch. Posteriorly, there was a large fluctuant swelling along the medial side of the popliteal fossa with extension into the calf. This popliteal synovial cyst was aspirated & 15 cc's of fluid was removed. Twenty-five cc's of fluid was removed from the suprapatellar fossa followed by a steroid preparation injection.

SLIDE 24 - TITLE: Scleroderma: Resorption of Mandible

CAPTION: This image shows absence of the angle of the jaw secondary to resorption of the mandibular condyle and ramus. This is a 56 year-old man with a seventeen-year history of Scl-70 (+) diffuse cutaneous systemic sclerosis. Resorption of the mandible may occur as a result of facial skin thickening with altered temporomandibular joint

function. Also evident in this photograph is significant wasting of the muscles of mastication--temporalis and masseter.

ADDITIONAL CLINICAL INFORMATION: At age 39, he presented with diffuse cutaneous systemic sclerosis with sclerodactyly, Raynaud's, and a bland myopathy with slightly elevated CK. Scl-70 antibody is positive. Tendon friction rubs are present over forearms and over anterior aspect of the ankles.

X-RAY DESCRIPTION: None

SLIDE 25 - TITLE: Smallpox induced premature fusion of several scattered epiphysis

CAPTION: 40 year old male who had smallpox during childhood presented with right lower limp pain. He had noticed shortening of right lower limp, finger. Patient reports that this started happening after smallpox occurred at the age of 6 years. Radiograph was suggestive of Smallpox induced premature fusion of several scattered epiphysis.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: X-ray

SLIDE 26 - TITLE: Pulmonary tuberculosis with multifocal tubercular osteomyelitis

CAPTION: 45 year old male presented B/L foot & right knee pain since 1 year. He had associated intermittent fever, weight loss. His investigations revealed sputum AFB positive, biopsy & culture from foot was consistent with tuberculosis.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: X-ray

SLIDE 27 - TITLE: Osteoarthritis: Sternoclavicular joint

CAPTION: A 77y/o female presented with gradual onset of pain and swelling on the medial side of the clavicles over the past 10 years associated with the bone growth. The pain initially started on her right now progressed on to her left clavicle. Diagnosis of osteoarthritis was made based on the clinical presentation and was confirmed with the CT imaging of the chest. Patient have Osteoarthritis in her other joints as well. Images A,B and C: Show the swelling in bilateral sternoclavicular joints in different angles. Image D: The CT Chest showing the Sternoclavicular joint and loss of joint space between the clavicle and the sternum, along with formation of the bone spurs. The findings consistent with arthritic changes.

ADDITIONAL CLINICAL INFORMATION: 77 y/o Caucasian female who is very active and carries all her daily activities of living on her own, was noticed to have gradual onset of pain and swelling on the medial side of the right clavicle 10 years ago at which time she was diagnosed with osteoarthritis of her right sternoclavicular joint. She presented to our clinic recently with the worsening of the swelling and she also started having similar findings on the medial side of her left clavicle. She was admitted and treated for pneumonia recently at which time CT chest with contrast was done, which clearly showed the findings consistent with osteoarthritis of her sternoclavicular joints.

X-RAY DESCRIPTION: CT chest - Chest with contrast Radiology report: March/2013, Osteoarthritis of bilateral Sternoclavicular joints. Left side worse than the right.

SLIDE 28 - TITLE: Psoriatic Arthritis of Hands and Feet and Hands X Rays

CAPTION: 60 years old male, diagnosed with Psoriasis 22 years ago, with no treatment. One year ago he started with polyarthralgias and edema on wrists, knees and ankles. low back pain and exacerbation of Psoriatic Plaques.

ADDITIONAL CLINICAL INFORMATION: Rheumatoid Factor (-), Anti CCP (-), ANA (-). due to economic limitations HLA B27 hasnt been performed on this patient.

X-RAY DESCRIPTION: X Rays: Right Hand: Multiple Bony erosions, destructive changes on First, Third and Fifth MCP, Third and Fourth PIP, Second and Third DIP. Left Hand: Multiple Bony Erosions, destructive changes of the First MCP and PIP. Third PIP.

SLIDE 29 - TITLE: Hemophilia Arthritis

CAPTION: 30 year old male with hemophilia and hemophilia related arthropathy.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: Radiograph(X-ray)Bilateral ankle radiograph demonstrates mild to moderate assymmetric joint space narrowing, large subchondral cysts, no bone demineralization, and minimal osteophytosis. There is abnormal angulation of the tibiotalar joint as a result of pediatric epiphyseal overgrowth; The epiphyseal overgrowth is related to episodes of early intraarticular hemorrhage.

SLIDE 30 - TITLE: DECT Scan of R hand showing scleroderma-related calcinosis of R-3rd extensor tendon and near ulnar head

CAPTION: 56 year old woman with longstanding diffuse scleroderma and extensive hand calcinosis

ADDITIONAL CLINICAL INFORMATION: painful, calcinosis deposit draining from soft tissue of the right hand.

X-RAY DESCRIPTION: DECT scan of R hand showing soft tissue calcinosis due to scleroderma along 3rd extensor tendon and near ulnar head.

SLIDE 31 - TITLE: Viral Synovitis in Child Detected by Ultrasound

CAPTION: Clinical History: A child, female of 5 years old, with monoarthritis in left knee, without pain, that began eight days ago and previously tonsillitis (2012/Aug/12). The 33clinical examination (2012/Aug/20): The child presented swelling left knee and performed ultrasound examination. The patient was been treated with anti-inflammatory no hormonal for one month. Diagnosis:Reactive arthritis viral.

ADDITIONAL CLINICAL INFORMATION: The exam laboratory (2012/Aug/23) showed: ANA negative; rheumatoid factor 53.0 U/ml; parvovirus B19 – Ig M 12.2 UI/ ml (normal reference: ≤12.1UI/ml); influenza IgM 1/320; ERS 26.0 mm; C3 125.0 mg/dl; C4 28.1mg/dl. Echocardiography and x-ray without alterations. After one month, the patient showed arthritis improvement and performed ultrasound follow - up.

X-RAY DESCRIPTION: Ultrasound exam (2012/Aug/20): Longitudinal lateral scan view of the left knee joint shows: hypoecic image by an important synovitis proliferation, with the flocculation aspect (star). Ultrasound exam (2012/Oct/15): Longitudinal lateral scan view of the left knee joint shows: without synovitis proliferation and isoecic image with fat tissue presence (arrow).

SLIDE 32 - TITLE: Adult Still's Disease: wrist deformities

CAPTION: PA view of the wrist demonstrate severe loss of articular cartilage of the radiocarpal, intercarpal and carpal metacarpal joints; fusion of the intercarpal and carpal metacarpal joints and osteopenia.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: x-ray

SLIDE 33 - TITLE: C1-C2 subluxation

CAPTION: 33 year old Hispanic female presents with primary C1-C2 subluxation as presenting symptom of rheumatoid arthritis after feeling numbness and tingling in her face and left shoulder for three months.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: MRI C spine T2 frFSE C2 dens irregularity with subluxation of the C1 ring associated with the dens, as well as circumferentially surrounding soft tissue, likely representing a pannus. This finding could be seen in connective tissue disease, such as rheumatoid arthritis

SLIDE 34 - TITLE: Rice bodies in the right knee of a patient with chronic, untreated seronegative RA

CAPTION: 37 y.o. white male was diagnosed with RA about 8 years ago, he couldn't afford the treatment, so he had been self-managing his arthritis with NSAIDs. The patient was found to have symmetrical, seronegative, erosive inflammatory arthritis with atlanto-odontoid articulation involvement as per C-spine imaging.

ADDITIONAL CLINICAL INFORMATION: The patient was c/o morning stiffness lasting for > 1hour, and arthralgia, the symptoms were mainly in the knees, elbows, wrists, PIP's and the neck. He was found to have evidence of synovitis on exam involving the knees, elbows, wrists and few bilateral hand PIP joints. Joint survey showed evidence of erosive arthritis with C1-C2 involvement.

X-RAY DESCRIPTION: Sagittal,proton density weighted image of the right knee without the use of IV contrast. RIGHT KNEE: 1. EXTENSIVE SYNOVIAL HYPERTROPHY AND PANNUS FORMATION WITH ASSOCIATED MARKED EROSION CHANGES ABOUT THE RIGHT KNEE AS DESCRIBED CONSISTENT WITH PATIENT'S HISTORY OF RHEUMATOID

ARTHRITIS. INNUMERABLE SMALL NONOSSIFIED BODIES WITHIN THE SUPRAPATELLAR POUCH REPRESENTING RICE BODIES/CHONDRAL BODIES CONSISTENT WITH SYNOVIAL CHONDROMATOSIS.

SLIDE 35 - TITLE: Scapulothoracic Bursitis

CAPTION: 22 yo Hispanic female diagnosed with systemic lupus erythematosus at age 7 and Sjogrens presenting with rashes, arthritis, malar rash, fatigue, raynauds, photophobia, dry eyes, vasculitic rash, chronically low C4 and proteinuria, +ANA, +SSA, and +SSB. She has previously been treated with plaquenil, prednisone and methotrexate. At the time of presentation to the rheumatology clinic she was not on any medications. Approximately 2 years ago she began to notice a mass at her right scapula that continued to slowly grow.

ADDITIONAL CLINICAL INFORMATION: MRI done to further evaluate this mass on her right scapula and thought to be a scapulothoracic bursitis that was eventually drained.

X-RAY DESCRIPTION: MRI chest-Multiplanar, multisequence MRI examination of the chest was obtained before and after the uneventful administration of 10 cc of MultiHance on a 1.5 GE scanner. Sequences obtained include coronal T2 SSFSE, axial T2 SSFSE with breath hold, axial T1 in and out-of-phase, axial firm non-breath hold, axial 2D FIESTA with fat sat, coronal T1, sagittal STIR, coronal STIR, sagittal T1, coronal LAVA precontrast with breath hold, axial LAVA precontrast with breath hold, axial LAVA postcontrast with breath hold, coronal LAVA postcontrast with breath hold, sagittal LAVA postcontrast without. A predominantly cystic lesion is again seen in the right posterolateral chest wall, deep to the serratus anterior muscle, located just inferior to the caudal tip of the right scapula. The lesion measures approximately 13.6 x 8 cm x 2.5 cm. Given differences in technique, this is slightly larger compared to the prior study dated 12/02/2011. This lesion is hypointense to skeletal muscle on T1 weighted imaging, and hyperintense on T2 weighted imaging. This lesion demonstrates peripheral, nodular enhancement after the administration of contrast. This mass does not have any extension to the ribs or adjacent lung parenchyma. In this area, findings are most consistent with bursitis of the adventitial scapulothoracic bursa. The nodular enhancement likely reflects a component of synovial hypertrophy

SLIDE 36 - TITLE: Gout - Arthroscopic Gross View Of Crystal Covering Knee Cartilage

CAPTION: 32 year old man underwent arthroscopic knee surgery due to anterior cruciate ligament injury during exercise. This picture was taken at the right knee as incidental finding, which suggested crystal disease. Also he had a history of some gout attacks. Polarized microscopic exam confirmed monosodium urate crystal.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: arthroscopic picture, knee

SLIDE 38 - TITLE: Scleroderma:Sclerodactyly with severe Raynaud's of hands

CAPTION: Male patient in his 20s, history of scleroderma and severe Raynaud's. In this photograph there is localized tightness of the skin of the fingers with ulceration. Together with poor circulation from Raynaud's disease this led to exposed bone, gangrene and auto-amputation of fingers.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: None

SLIDE 39 - TITLE: Heliotrope rash and periorbital edema in dermatomyositis

CAPTION: Violaceous rash on the eyelids accompanied by eyelid and periorbital swelling in a 67 Year-Old Caucasian male with hypomyopathic dermatomyositis.

ADDITIONAL CLINICAL INFORMATION: Presented with raised red rash over the face and dorsum of the hands which then extended to the chest, shoulders, back, neck, and scalp in few weeks; also with swelling around the eyes since the appearance of the rash.

X-RAY DESCRIPTION: Heliotrope rash, periorbital edema, dermatomyositis

SLIDE 40 - TITLE: Cocaine Vasculitis

CAPTION: A 27 year old male with a history of nasal cocaine use was admitted after 8 months of progressive nasal edema, rhinorrhea, crusting, and erosion into his upper lip. Prior to this, the patient was in apparent good health other than the start of cocaine use one year prior to admission.

ADDITIONAL CLINICAL INFORMATION: Shown is the midline destructive lesion involving his upper lip and the nasal cavity with progression to erosion of the hard palate due to continued cocaine use. A month after presentation, he also developed petechial lesions on his feet (image shown) that resolved over next 2 weeks. Upon admission, the patient underwent nasal biopsy, which showed acute and chronic inflammation and no granulomas. He was c-ANCA positive in 1:640 titer and anti-PR3 positive. ANA, RF and p-ANCA were negative. He had elevated ESR at 60. His urinalysis was normal and chest CT scan did not reveal cavitary or granulomatous lesions. Patient's differentials included cocaine-induced vasculitis vs granulomatous polyangiitis with the former favored due to lack of renal and pulmonary involvement. We planned to check anti-neutrophil elastase antibodies but the patient was lost to follow-up.

X-RAY DESCRIPTION: None

SLIDE 41, 42, 43, 44

#1 - CASE TITLE: The Many Faces of Cocaine-Induced Vasculitis

CASE DESCRIPTION: Levamisole-induced vasculitis with features of granulomatosis with polyangiitis (GPA). A 30 year old Caucasian female presented to the emergency room with worsening of chronic skin ulcerations following longstanding cocaine abuse and stridor. Dermatological exam revealed multiple eschar-like lesions in various stages of healing on the extremities, specifically the lateral knees, thighs, and multiple areas on the hands and digits (Figure 1). Admission laboratory findings revealed an elevated creatinine of 5.4 mg/dl (reference range 0.5-1.2); six months prior creatinine had been normal at 0.8 mg/dl. CBC showed leukopenia with a WBC count of 2,200/mm³ (reference, 4,000-10,000) with a differential of 62.6% granulocytes (reference, 50-70), 25.6% lymphocytes (reference, 20-40), 11.8% monocytes (reference, 2-8), 0% eosinophils (reference, 0-5), and 0.6% basophils (reference, 0-1). Bone marrow biopsy revealed hypoplastic marrow with negative cultures and no evidence for malignancy. FANA was positive >1:640 (homogenous pattern). dsDNA, SSA, SSA and Smith antibodies were negative. Hepatitis C antibody was positive; however, cryoglobulins were negative. Hepatitis B serology and HIV were negative. Urine drug screen revealed cocaine. Urinalysis showed 4+ hematuria and red blood cell casts. C-ANCA (directed to anti-proteinase 3 antibody), P-ANCA directed to anti-myeloperoxidase antibody and anti-neutrophil elastase antibodies were all positive with titers of 13.4 U/ml (reference, 0-3.5), 15.9 U/ml (reference, 0-3.5) and 1.059 O.D (reference < .25 O.D) respectively. Laryngoscopy showed edema of the trachea with no obstructive lesions identified. A renal biopsy revealed focal necrotizing and crescentic glomerulonephritis consistent with pauci-immune ANCA associated vasculitis (Figure 2). A maxillofacial CT displayed paranasal sinus disease along with anterior nasal septal defects (Figure 3). CT of the chest revealed multiple noncalcified nodules in each lung lobe with no evidence of lymph node enlargement (Figure 4). Based on clinical and laboratory findings, a diagnosis of vasculitis secondary to cocaine abuse was made. The patient required a tracheostomy but did respond to high-dose steroids and rituximab with normalization of the renal function.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: Maxillofacial CT displayed paranasal sinus disease along with anterior nasal septal defects. Chest CT revealed multiple noncalcified nodules in each lung lobe with no evidence of lymph node enlargement.

HISTOPATHOLOGY DESCRIPTION: Renal biopsy using H&E X200 revealed focal necrotizing and crescentic glomerulonephritis, pauci-immune type consistent with ANCA-associated glomerulonephritis. Images show fibrous crescent formation.

SLIDE 45, 46

#2 - CASE TITLE: SLE related spontaneous tendon rupture

CASE DESCRIPTION: HPI: 35 year old African American Female presents to ER with excruciating left knee pain Her PMH is significant for systemic lupus erythematosus (SLE) with speckled high titer ANA, malar rash, arthritis, oral ulcers, serositis, low complements, lupus cerebritis, bilateral spontaneous rupture of patellar tendons – Left while walking on even surface, Right one day later with snap and pain. PSH: hand, ankle, nasal surgeries; knee surgeries for spontaneous tendon rupture. She had immediate operation on the knees without success followed by failed revision. She was deemed not to be a surgical candidate for farther knee repairs after those surgeries. Medications for lupus: prednisone 40 mg a day and hydroxychloroquine 200 mg twice daily Pertinent musculoskeletal physical exam: L hand with severely contracted fingers, not tender, no warm, not boggy, R hand less contracted with scars on fingers, R

thumb with swan neck deformity that is reducible, not allowing to roll L hip, R hip rolling fine, R knee with scar and non tender, L knee with patella that is displaced laterally and cephalic, there is fluid and warmth and it is very tender to palpation

ADDITIONAL CLINICAL INFORMATION: Assessment, plan of care and treatment Acute inflammation of the left knee with mild lupus flare; patient on immunosuppressive so cannot rule septic arthritis of the left knee Knee anatomy made arthrocentesis technically difficult, but 15 cc yellow fluid aspirated and analysis revealed non –infected, inflammatory fluid. The knee was injected with steroids. Patient was discharged home a day later. Image case summary Rheumatologist should be aware of spontaneous tendon rupture as a rare complication of SLE and adjust the technique of joint aspiration and management accordingly. Literature review suggests that female gender and use of steroids are risk factors for this condition. The knee is the site of the rupture in more than 60% of cases. Trauma is not responsible for the injuries in the cases analyzed and immediate surgical repair is the primary treatment of the rupture in that location (1). Another study suggests that Jaccoud's arthropathy is present in 29 % of the cases of spontaneous tendon rupture (2). references: 1. Tendon rupture in systemic lupus erythematosus. Semin. Arthritis Rheum.1988 Nov;18(2):127-33. Furie, Chartash 2. Spontaneous tendon rupture in systemic lupus erythematosus: Association with Jaccoud's arthropathy. Lupus (2010) 19, 247–254. Alves, Macieira

X-RAY DESCRIPTION: XR KNEE LEFT AP & LATERAL No acute fracture. There is an evidence of patellar tendon rupture with superior displacement of the patella and fluid and loose bodies inferior to the patella distending Hoffa's fat pad. Old injury to the lateral tibial plateau is seen, with depression. Stable serpiginous opacities in the distal femoral metaphysis and proximal tibial metaphysis are favored to represent bone infarcts. Stable tubular ossification medial to the distal femur may represent heterotopic ossification.

SLIDES 47, 48, 49 50

#3 - CASE TITLE: Drug-induced Sweet's Syndrome Masquerading as ANCA-associated Vasculitis

CASE DESCRIPTION: A 59 year-old male with history of myelodysplastic syndrome developed acute renal failure, hematuria, proteinuria, hypoxia, pulmonary infiltrates, and skin rash 6 days after receiving GM-CSF (Neulasta) and was diagnosed with Drug-induced Sweet's Syndrome with cutaneous and extracutaneous features.

ADDITIONAL CLINICAL INFORMATION: A 59 year-old male with congenital solitary kidney and squamous cell cancer of the lung complicated by myelodysplastic syndrome was admitted with fevers and cough from a long term care facility. He was started on broad spectrum antibiotics to treat healthcare associated pneumonia. His respiratory status initially improved with antibiotics. One dose of GM-CSF (Neulasta) was given for significant neutropenia. Six days after his Neulasta dose, the patient was noted to have worsening anemia, new onset hematuria and proteinuria, acute renal failure (creatinine increase from 1.2 to 5.0), and worsening respiratory status (hemoptysis, hypoxia). He also developed a purpuric rash on hands, forearms, and left thigh. Laboratory workup revealed elevated inflammatory markers, negative infectious workup, positive ANA 1:160, and high titer myeloperoxidase and proteinase-3 antibodies. The primary service was initially concerned for ANCA-associated vasculitis with pulmonary, renal, and skin involvement. Skin biopsy of the rash showed a dermal neutrophilic infiltrate consistent with febrile neutrophilic dermatosis. Chest CT showed diffuse alveolar infiltrates and bronchoscopy showed nodules in the mainstem bronchi consistent with tracheobronchial Sweet's syndrome. A diagnosis of Sweet's syndrome with renal and pulmonary involvement was made and the patient's rash and respiratory status improved with high dose steroids. Unfortunately his renal function did not recover to baseline.

X-RAY DESCRIPTION: chest CT without contrast - development of new coalescing alveolar infiltrates throughout the right lower lobe and patchy nodular infiltrates scattered throughout the left lung along with new small left pleural effusion.

HISTOPATHOLOGY DESCRIPTION: 4 mm punch biopsy, left thenar eminence, low and high power views, hematoxylin and eosin stain: Dermal neutrophilic infiltrate with extravasated red blood cells. PAS and Gram stains negative for microorganisms. Sections show acral skin with focal epidermal spongiosis. Within the dermis is a sharply localized collection of neutrophils and neutrophilic debris admixed with extravasated red blood cells. No fibrinoid necrosis of vessels is seen. No eosinophils are appreciated.

SLIDE 51, 52, 53,54

#4 - CASE TITLE: Hypertrophic Pulmonary Osteoarthropathy

CASE DESCRIPTION: A 63-yo African American male presented with progressive SOB for weeks, weight loss, fevers, chills and on CT scan of chest was found to have a large right hilar mass measuring 6.6 x 5.2 x 6.1 cm. Biopsy of the mass showed non-small cell lung cancer. Rheumatology consult called for management of diffuse body pains.

ADDITIONAL CLINICAL INFORMATION: He c/o pain in bilateral wrists, hands, feet and legs for past 2 weeks. No prior history of joint pains. The pain gets worse by pressing on the involved sites and better with rest or elevation. Musculoskeletal exam showed clubbing of fingers and toes bilaterally, edema + diffusely over hands, legs and feet. Very tender to palpation over b/l wrists, MCP/PIP/DIPs, elbows, anterior tibial surfaces, ankles.

X-RAY DESCRIPTION: 1) X-Ray of hands and wrists: There is moderate dorsal soft tissue swelling over the metacarpals bilaterally. Periosteal reaction is noted along the bilateral fourth and fifth metacarpals and to a lesser degree the left third metacarpal. There is no acute fracture or dislocation. The joint spaces are maintained. No erosions are identified. Periosteal reaction is noted along the radius and ulna bilaterally. 2) Whole body bone scan: Following IV injection of 20 mCi 99mTc-MDP, planar images from the skull vertex to both lower extremities were obtained approximately 3 hours post injection. Impression: Diffuse periosteal reaction along the bilateral femurs, tibia and forearms, consistent with hypertrophic pulmonary osteoarthropathy secondary to right infrahilar lung mass seen on CT chest. No focal lesions to suggest metastatic disease to bone.

SLIDE 55, 56

#6 - CASE TITLE: Acromegaly

CASE DESCRIPTION: A 47 year old Indian man presented in June 2011 with left knee swelling and discomfort.

Examination showed a large left knee effusion. Some synovial swelling of the PIP joints was noted on examination, but the patient had no joint symptoms except for his left knee. Aspiration of the knee yielded non-inflammatory fluid. No crystals were identified. An intra-articular corticosteroid injection provided relief. ESR, rheumatoid factor, ANA, Lyme ELISA were negative.

ADDITIONAL CLINICAL INFORMATION: Patient had knee swelling and discomfort in June 2011. Aspiration yielded non-inflammatory synovial fluid with no crystals seen on examination. ANA, RA factor, ESR, were negative. Left knee x-rays showed mild medial joint space narrowing and small osteophytes. Knee symptoms improved with corticosteroid injection. He had repeat aspiration and injection of his knee in January 2012. In November 2012, he developed acute onset of swelling and pain of his right third MCP joint. He also complained of mild recurrent left knee swelling. He was treated with a short course of oral prednisone and further studies were done including ANA, RA factor, CCP antibodies, ESR and CRP all of which were normal. Hand x-rays showed prominent spade like phalangeal tufting and soft tissue swelling around the PIP joints. A screening IGF-1 level was significantly elevated and he was found to have a growth hormone producing pituitary macroadenoma. On further questioning, he admitted to enlarging ring size over the last 6 years and coarsening of facial features noted by family on a visit to India. He also had hypertension and a history of snoring. He underwent successful resection of the macroadenoma with resolution of joint symptoms.

X-RAY DESCRIPTION: X-rays of left knee show medial joint space narrowing with small osteophytes X-rays of hands (bilateral) show prominent phalangeal tufting, soft tissue swelling around PIP joints, widening of MCP joint spaces T1 MRI shows a 1.5 cm mass in the pituitary

SLIDE 57,58,59,60

#7 - CASE TITLE: Type IV Ehlers-Danlos Syndrome in a 49-year-old male with renal infarct

CASE DESCRIPTION: Image 1 shows left rectus sheath hematoma (arrow) in a 49-year-old male with Ehlers-Danlos syndrome, type IV (vascular EDS). Image 2 shows multiple renal infarcts (arrow) for which he presented with severe left-sided abdominal pain and markedly elevated blood pressure (BP 211/168). CT angiogram (Images 3 and 4) shows bilateral renal artery fusiform dilatations/aneurysms (arrows), without wall thickening. Image 4 also shows that he has two right renal arteries and a single left renal artery. The right renal artery supplying the right mid and upper pole and the mid portion of the left renal artery demonstrate fusiform dilatation (arrows).

ADDITIONAL CLINICAL INFORMATION: Image number 1: Seven months prior to his evaluation at our rheumatology clinic, Mr. A noticed left-sided abdominal pain after repetitive weight lifting. He was seen at an outside hospital. His hematocrit dropped significantly for which he required 4 units blood transfusion. He did not have signs of other bleeding. His hematocrit remained stable and on the follow up images, the left rectus sheath hematoma was almost all absorbed. Image number 2-4: About one month prior to his evaluation, Mr. A experienced severe sudden onset left-sided abdominal pain. He did not have clinical signs of infection: he did not have fever and his urine analysis showed 2+ blood without white blood cells. He had negative blood and urine culture. CT scan with contrast at that time showed resolved left rectus sheath hematoma and multiple levels of low attenuations in a somewhat triangular or wedge-shaped configuration distribution. The images are consistent with renal infarcts. CT angiogram showed fusiform dilatation of the bilateral renal arteries without wall thickening. Further work up found he has a pathogenic mutation in COL3A1 gene, which is known to be associated with type IV Ehlers-Danlos syndrome (Hum Mutat. 1997. 9 (5): 475).

X-RAY DESCRIPTION: Image 1: CT without contrast, axial image shows a large round mainly isodense mass (arrow) posterior to the rectus abdominis muscle. This is consistent with a big left rectus sheath hematoma Image 2: CT with contrast, axial image shows multiple isolated areas of low attenuation (arrow) in a somewhat triangular or wedge-shaped configuration distribution. This is consistent with renal infarct. Image 3: CT angiogram, axial image shows bilateral renal arteries aneurysmal dilatation (arrows), measuring up to 14 mm in diameter. Image 4: reformatted CT angiogram: coronal view shows two right renal arteries and a single left renal artery. The origins of all three renal arteries appear normal. The right renal artery supplying the right inferior pole is unremarkable. The right renal artery supplying the right mid and upper pole and the mid portion of the left renal artery demonstrate fusiform dilatation (arrows)

SLIDE 61, 62, 63, 64, 65,66

#8 - CASE TITLE: Hereditary Multiple Exostoses

CASE DESCRIPTION: Case: 50 year old African American female with history of familial osteochondromatosis presented with multiple joint pains in her hands, bilateral hip and knees. On physical exam: Non tender palpable bony masses in the wrists, groin area and knees bilaterally. Bilateral knee joint line tenderness. Painful and limited range of motion of her hips and knees bilaterally.

ADDITIONAL CLINICAL INFORMATION: *Hereditary Multiple Exostoses (HME)* also known as Diaphyseal aclasis or multiple osteochondromatosis is characterized by the development of two or more osteochondromas, growing outward from the metaphyses of long bones. Most cases are caused by autosomal dominant inheritance of a germline mutation in the tumor suppressor genes, exostosin EXT1 or EXT2. However, spontaneous mutations also occur.

X-RAY DESCRIPTION: Imaging: 1. Plain radiographs of the hand, pelvis and knees (Figure 1, 2,3,4,5,6,7) showed multiple osteochondromas. 2. CT pelvis: Numerous exostotic lesions right and left iliac crest, right acetabulum, left greater trochanter (Figure 8,9) 3. MRI knees: Osteochondromatosis of the distal femur and proximal tibia with prominent cartilage cap of the medial tibial metaphyseal osteochondroma (Figure 10,11)

SLIDE 67,68

#9 - CASE TITLE: Cryoglobulinemia

CASE DESCRIPTION: Age: 62 years Female two years ago was admitted for leg pain, and paresthesias On physical examination we found arthritis of wrist, metacarpophalangeal joints on both hands, and feet with purpuric lesions, hypothermia absence of pedal and popliteal pulses. Her ankle/braquial index was 0.9 (initially we considered atherosclerotic disease) Her rheumatoid factor was 56 U/I, CRP 5.6 mg/dl, and cryoglobulins positives (3+); HBV, HCV, ACL, LA, and ANA, negatives. The angiotomography evidenced absence of circulation of both tibial and pedal arteries. Antibiotics, anticoagulation and prednisone were started. Skin biopsy revealed leukocytoclastic vasculitis. Because she decided voluntary hospital discharge, we couldn't classified the type of cryoglobulins Diagnosis: Cryoglobulinemia

ADDITIONAL CLINICAL INFORMATION: Disease duration: 2 years Serologies: antinuclear antibodies negative, rheumatoid factor: 56 U/I (range: 0-32 U/I), cryoglobulins positive (3+), Hepatitis B and C negative, HIV negative

X-RAY DESCRIPTION: Contrasted angiogram. Reduced circulation of right anterior and posterior tibial arteries; lack of circulation on left anterior, posterior tibial and popliteal arteries

SLIDE 69

#10 - CASE TITLE: Cryoglobulinemic Vasculitis

CASE DESCRIPTION : 34 yo Hispanic woman with history of SLE (arthritis, malar, serositis, Lupus nephritis class III, DNA, Smith, ANA, vasculitis,) and asymptomatic Ro + presents with pleuritic chest pain and was empirically treated with prednisone 20mg daily.

ADDITIONAL CLINICAL INFORMATION: 3 days later develops petechial and hemorrhagic areas of the tongue followed by cutaneous palpable purpura of the Legs.

10 days later she develops nodular lesions on the palms of the hands that were intermittent but very painful with residual burning sensations. Palms remained erythematous throughout the patient episode of vasculitis. She in addition developed gross arthritis of the bilateral knees, ankles, MCPs and elbows.

X-RAY DESCRIPTION: None

HISTOPATHOLOGY DESCRIPTION: Patient was noted to have high DNA, low complements, elevated immunoglobulins and high Total protein, low wbc, Hg 10 and elevated ESR and CRP. UA was significant for Blood and protein.

SLIDE 70, 71

#11 - CASE TITLE: Scleredema of Buschke

CASE DESCRIPTION: 46 yr old African American male with PMH of DM-1, diabetic nephropathy and retinopathy presented to the clinic for the evaluation of thickened skin on his back and upper shoulders. On physical exam his skin was thickened, woody and indurated from his neck down to his lower back extending all across the trunk and over the shoulders and posterior aspect of upper arms. Skin biopsy of the upper back revealed thickened dermis, fenestrated collagen and mucin deposition in deep dermis, consistent with scleredema.

ADDITIONAL CLINICAL INFORMATION: His medical history was significant for type 1 Diabetes Mellitus, complicated by retinopathy and nephropathy. Review of systems negative for raynauds, phenomenon, dysphagia, dyspnea or joint pain. Alert and oriented without any distress. SKIN: Thickened and indurated skin from the neck down to the lower back extending all across the trunk and over the shoulders and posterior aspect of upper arms. Normal skin of the anterior chest wall, hands and feet.

X-RAY DESCRIPTION: None

HISTOPATHOLOGY DESCRIPTION: Skin biopsy of the upper back revealed thickened dermis, fenestrated collagen and mucin deposition in deep dermis, consistent with scleredema.

SLIDE 72

#12 - CASE TITLE: Resolution of Soft Tissue Calcification in CREST

CASE DESCRIPTION: 33 year old female with CREST syndrome diagnosed after these images were obtained showing extensive calcification in the soft tissues of the lateral hips. She was started on a calcium channel blocker which she did not take regularly but over the following years shown in images taken two and six years later, she had near resolution of the calcifications.

ADDITIONAL CLINICAL INFORMATION: Initially had Raynaud's primarily with negative ANA. Carried diagnosis of undifferentiated connective tissue disorder initially.

X-RAY DESCRIPTION: X-ray

SLIDE 73, 74,75,76

#13 - CASE TITLE: Sarcoid arthropathy

CASE DESCRIPTION: 56 year old black male presented to the rheumatology clinic for evaluation of hand pain and swelling which started 5 years ago and has worsened gradually (image 1).

ADDITIONAL CLINICAL INFORMATION: Erythrocyte sedimentation rate was 12mm/hr (ref 0-13mm/hr) and c-reactive protein 5.5mg/dL (ref < 7mg/dL). Complete blood count and basic metabolic panel were within normal limits. Rheumatoid factor, anti-CCP and antinuclear antibodies were negative. Biopsy of a cervical lymph node demonstrated non-caseating granulomas consistent with sarcoidosis.

X-RAY DESCRIPTION: Hand films were notable for demineralization with a "lace-like" appearance of bilateral 4th middle phalanges, bilateral "hook-like" osteophytes, bilateral periostitis, bilateral clubbing and left wrist scapholunate advanced collapse (SLAC wrist) (images 2 and 3). Chest CT was notable for calcified mediastinal and hilar lymphadenopathy (image 4).

SLIDE 77

#14 - CASE TITLE: Chronic Non Bacterial Osteomyelitis (CNO)

CASE DESCRIPTION: 16 yr old F with Left clavicular swelling

ADDITIONAL CLINICAL INFORMATION: 16 yr old F presented to pediatric rheumatology clinic for a 4 yr history of L clavicular swelling. She had an extensive work up outside including X-ray, CT, Bone biopsy of L clavicle to rule out initial suspicion for bone tumor, Ewing's sarcoma, lymphoma, infectious osteomyelitis. Imaging and path was reviewed and repeated. Time course, location and negative cultures were supportive of a diagnosis of sterile immune mediated bone inflammation consistent with CNO which is a milder subset of Chronic recurring multifocal osteomyelitis (CRMO). The patient responded very well to treatment with NSAIDs. MSK Exam: Marked bony enlargement of the left clavicle from the midline to about 2/3 of the length laterally; mildly painful on palpation and minimally warm to touch. ROM is full in all joints. No evidence of enthesitis. Pertinent positive Labs: ESR: 72, c-RP: 1.4mg/dL. She underwent an extensive work up at outside hospital, including imaging and bone biopsy to rule out malignancy: Ewing's sarcoma, lymphoma, infectious osteomyelitis. Outside imaging was reviewed and repeated including bone biopsy from left clavicle

X-RAY DESCRIPTION: X-ray left clavicle: enlargement of the left clavicle with increased sclerotic and mildly lytic foci involving the proximal and mid part of the left clavicle.

CT left clavicle without contrast: expansile lesion involving most of the left clavicle with mixed sclerotic and lytic areas. Bone Scan: increased uptake in the left clavicle. Left clavicle, core biopsy: Viable lamellar bone with paucicellular marrow with fibrosis, consistent with chronic osteomyelitis. No infectious etiology or malignancy was identified after a thorough evaluation.

SLIDE 78,79

#15 - CASE TITLE: Keratoderma Blennorrhagicum

CASE DESCRIPTION: A 19 year old male presented with a 4 month history of rash and arthritis. This annular, erythematous, non pruritic, non painful rash was located on the palms, soles and knees. He reported debilitating joint pain, swelling in the knees, wrists and ankles and a 50lb weight loss over 4 months. Three months prior to admission, he had been diagnosed and treated for gonorrheal and chlamydial urethritis. Admission physical exam revealed temperature of 102F, mild synovitis in the wrists, moderate synovitis in the ankles, large bilateral knee effusions and achilles tenderness and swelling. His skin exam showed scaling annular plaques on the extensor surface of the knees bilaterally (Panel A), scaling plaques near the glans penis, and erythematous scaling papules and plaques on the left palm. He had diffuse hyperkeratotic nodules on the plantar aspect of both feet (Panel B) and onycholysis of the nails, nail pitting and subungual keratosis.

ADDITIONAL CLINICAL INFORMATION: His labs revealed HLA-B27 positivity, ESR of 116 mm/hr, and CRP of 17.12 mg/dl. Synovial fluid obtained from left knee showed an inflammatory fluid with 27891 nucleated cells per cubic millimeter, 83% neutrophils, no crystals and culture negative for bacteria. Punch biopsy of annular lesions on the left knee revealed neutrophilic microabscesses, elongation of the rete pegs, and confluent parakeratosis consistent with psoriasiform dermatitis (Panel C). The patient was diagnosed with reactive arthritis and started on Prednisone 60 mg daily and topical corticosteroids to be used on feet, palms, knees. He improved and was discharged home.

X-RAY DESCRIPTION: None

HISTOPATHOLOGY DESCRIPTION: Hematoxylin-Eosin stain, Punch biopsy of annular lesions on the left knee revealed neutrophilic microabscesses, elongation of the rete pegs, and confluent parakeratosis consistent with psoriasiform dermatitis

SLIDE 80

#16 - CASE TITLE: Pyogenic arthritis, pyoderma gangrenosum, and acne (PAPA) syndrome: skin manifestations

CASE DESCRIPTION: This is a 26 year old Caucasian man with pyogenic arthritis, pyoderma gangrenosum, and acne (PAPA) syndrome. The picture demonstrates several phases of pyoderma gangrenosum, progressively healing with high doses of steroids and infliximab. PAPA is an autosomal dominant syndrome of recurrent pyogenic arthritis, pyoderma gangrenosum, and acne. Patients typically present early in childhood with episodes of sterile monoarticular arthritis, and later in life develop pyoderma gangrenosum-like ulcerative skin lesions and cystic acne. Activation of the interleukin 1 β pathway with IL 1 β overproduction play central role in the pathogenesis and in addition TNF- α levels are elevated in the mononuclear cells of patients with PAPA syndrome. Currently the patient is maintained on a combination of therapy with anakinra and infliximab with occasional relapses of his lesions.

ADDITIONAL CLINICAL INFORMATION: None

X-RAY DESCRIPTION: None

SLIDE 81,82

#17 - CASE TITLE: Presumed IgA Vasculitis

CASE DESCRIPTION: 58 y/o Caucasian male with no significant past medical history.

ADDITIONAL CLINICAL INFORMATION: He initially presented to on 5/29/13 with rash on the lower extremities x 1 day, diffuse myalgias (mild) x 1 day, left testicular pain (dull, 1/10) x 1 day, and lower abdominal pain (colicky, mild) x 2 day. His ANA and ANCA were negative; ESR=24 and CRP=2.98. He underwent punch biopsy of the skin that showed small vessel vasculitis (leukocytoclastic vasculitis) with DIF negative for IgA. He was discharged home on 6/1/13. He had worsening of the abdominal pain and an episode of melena for which underwent EGD on 6/4/2013 that showed diffuse gastric petechiae and severe duodenal ulcers (in the 2nd and 3rd parts of duodenum). He was started on prednisone 20 mg b.i.d. His CT abd/pelvis with PO contrast showed severe bowel wall thickening with surrounding inflammatory stranding starting in the 3rd portion of the duodenum through the proximal jejunum. Urine microscopy: 5 RBCs; Pr/Cr=0.2. He was started on Solu-MEDROL 40 mg IV daily and protonix 40 mg IV b.i.d. Rheumatology team was called on 6/7/13. Our initial main differential included IgA vasculitis and polyarteritis nodosa. His glucocorticoid dose was increased to IV Solu-MEDROL 50 mg b.i.d. His CBC showed a neutrophilic leukocytosis; WBC of 13K and his CMP revealed an albumin of 2.8; otherwise unremarkable with Cr=0.9. He had low C3: 78 and low C4:<10 but his other autoimmune serologies were unremarkable including negative RF, anti-Smith, RNP, SSA, SSB, cryoglobulins. His SPEP with no monoclonal proteins. His serum IgA level was normal. His other tests were unremarkable including negative LAC, ACL, and beta₂ GPI, Hep B and C serologies, HCV PCR, HIV, blood cultures, PPD. Repeat skin punch biopsy on 6/7/13 showed perivascular deposits of IgA, fibrin, C3, IgM, and IgG in the superficial and mid dermis which was most suggestive of IgA vasculitis but unusually the deposits also involve medium vessels which is not characteristic of this process. He underwent abdominal MRA on 6/8/13 that showed mild irregularity of both renal arteries as well as mild beading and irregularity of the distal superior mesenteric artery and proximal several jejunal branches, and irregularity of the splenic artery suggestive of vasculitis. He underwent angiogram (celiac, SMA, renal arteries) on 6/11/13 that was normal. He was discharged on 6/12/13 on prednisone 50 mg b.i.d. (1 mg/kg/day). He had a follow up on 6/25/13 for presumed IgA vasculitis. He was clinically stable and no clinical features of active systemic vasculitis. Repeat BMP, UA, urine Pr/Cr were stable. He was advised to taper his prednisone dose by 10 mg weekly.

X-RAY DESCRIPTION: MRA abdomen with IV contrast Angiogram (celiac, superior mesenteric artery, renal arteries)

HISTOPATHOLOGY DESCRIPTION: Skin punch biopsy: perivascular deposits of IgA, fibrin, C3, IgM, and IgG in the superficial and mid dermis.