SIX ILLUSTRATED RHEUMATOLOGY CASES

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1] Osteopoikilosis

A 23-year-old male presented with bilateral knee selling. He had history of similar problem One and half year ago treated with joint aspiration and non-steroidal anti-inflammatory drugs. There was neither history of bowel or genitourinary complaints nor any significant family history. Clinical examination revealed bilateral knee arthritis and enthesitis on medial side of left ankle suggestive of spondarthritis. General examination did not reveal any abnormalities. Rheumatoid factor was normal, ESR was 41 mm/1 hr and synovial fluid was inflammatory. His X-ray revealed Osteopoikilosis (Fig. 1).

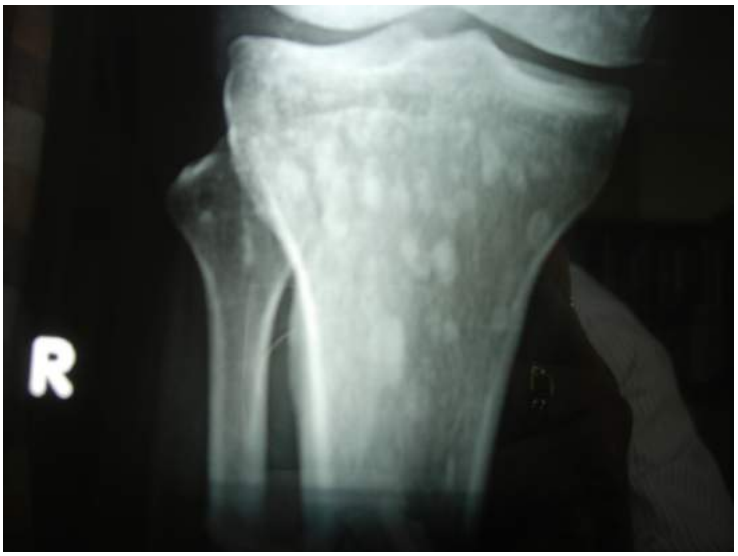

Fig 1. Osteopoikilosis (Right knee Joint)

Osteopoikilosis (also known as spotted bone disease) is an autosomal dominant condition characterized by multiple, symmetric, pea-sized round or oval discrete sclerotic densities usually situated near epiphyses and adjacent part of metaphyses. Skull, ribs and vertebrae are not involved in this condition. These densities are

multifocal compact bony islands which appear in childhood and persist throughout life. Buschke-Ollendorff syndrome is osteopoikilosis associated with skin lesions. This benign asymptomatic condition does not require any further investigations or treatment.

2] Lofgren's Syndrome (Acute Sarcoid Arthritis)

58-year-old male, a retired bank employee, presented with 3-week history of bilateral ankle pain and swelling. He also had intermittent dry cough that would be continuous on lying down. 2 painless red nodules developed recently on his left lower extremity. He had recent onset fatigue and lost 2 Kg weight during those 3 weeks. He was a known hypertensive on drugs for 10 years and had skin and nail psoriasis for 34 years. Clinical examination revealed synovitis of both ankles and wrists. He had skin and nail psoriasis along with discrete lesions of erythema nodosum on legs and right hand. A few inspiratory crackles could be auscultated on respiratory examination. ESR was 57 mm at one-hour, Rheumatoid factor 57 IU and Serum ACE normal. Other laboratory investigations were normal. His X-ray of chest showed hilar prominence confirmed by CT scan to be lymphadenopathy. He was treated with a 4-week tapering course of prednisolone and settled completely without any recurrence for next 3 years.

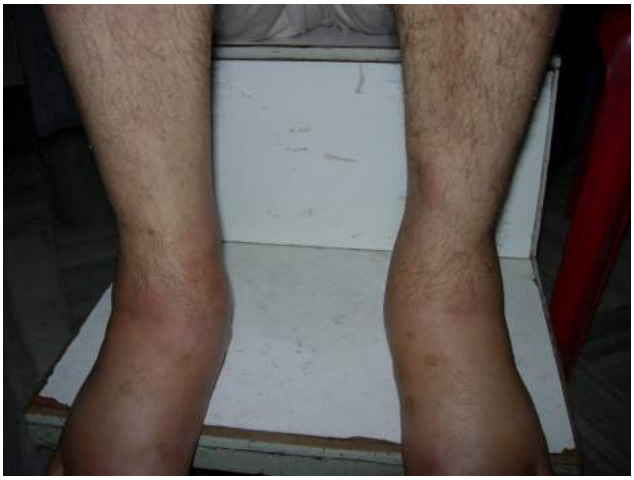

Fig 2. Erythema nodosum in Sarcoid Arthritis

Lofgren's syndrome requires 3 out of following 4 criteria for diagnosis:

1. Bilateral ankle arthritis
2. Duration less than 2mths
3. Erythema nodosum (Fig. 2)
4. Age less than 40yrs

Lofgren's syndrome is a clinical diagnosis and does not require biopsy evidence of sarcoidosis. Serum ACE is usually normal. Treatment comprises of NSAIDs and steroids and most cases recover within 4 weeks.6% cases can recur during next 20 years. Association with psoriasis appears to be incidental and rarely reported.

3] Osteitis condensans ilii

A 52-year-old female presented with intermittent low back pain of varying severity radiating to right thigh for 3 years. The pain was more frequent in cold season and increased after exertion. She underwent hysterectomy 12 years back for dysmenorrhoea. Laboratory investigations, including inflammatory markers, were

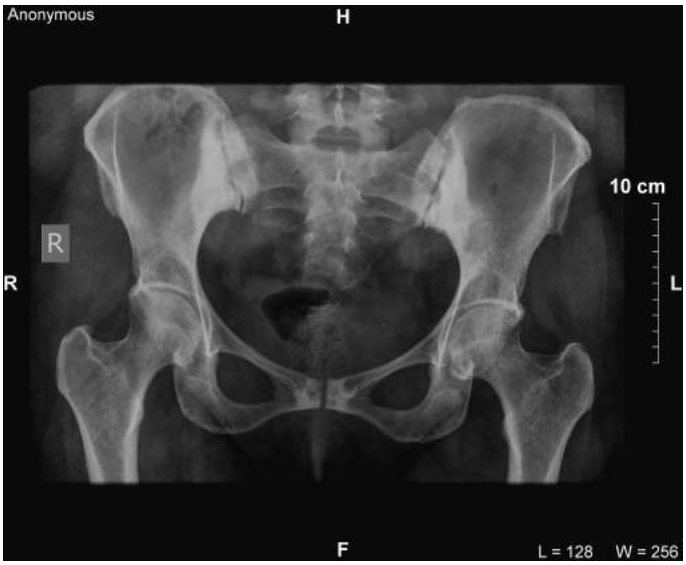

Fig 3. Osteitis condensans ilii

normal. Pelvis X-ray shows dense iliac sclerosis adjoining sacroiliac joints. T2 weighted axial MRI image shows well-defined hypo intense areas around both sacroiliac joints suggesting sclerosis (Fig. 3). No obvious erosions or effusion suggesting sacroiliitis were seen.

Osteitis condensans ilii is a benign condition more common in obese multiparous females. Low back pain radiating to buttocks, usually exacerbated by activity and relieved by rest, is a common feature. Pain is. Back pain may be milder in some cases. X-ray shows bilateral symmetric sclerosis on iliac sides of sacroiliac joints. The sclerosis is triangular in shape with base pointing inferiorly. Sacral sclerosis may accompany and the condition can be unilateral. Sacroiliitis can be differentiated by clinical examination and MRI in doubtful cases. Bone remodeling due to stress induced vascularity across sacroiliac joints is possible causative factor. Similar isolated changes can occur in pubic bones, medial end of clavicle and other sites. The condition may resolve spontaneously and radiological picture may also change from time to time. Therapy consists of posture correction and active exercises for 6-12 months or sacroiliac fusion surgery for nonresponsive cases.

4] Bisphosphonate induced Osteonecrosis of Jaw

48-year-old male, a case of ankylosing spondylitis, was receiving Ibandronic Acid (150 mg once a month) since September 2007. He developed a crack in his right molar tooth which was subsequently removed in April 2009. The wound did not heal and an extensive debridement was performed two and half months later. A CT scan in July 2009 showed a lytic area with cortical break in the right side of mandible in relation to the extracted molar teeth and another lytic area in right side of the body of mandible with no contact with the roots of the right sided teeth.

The incidence of osteonecrosis of jaw in patients on low dose amino-bisphosphonates is estimated to be 1 in 10000 to 1 in 100000. The exact cause-effect relationship is not yet established.

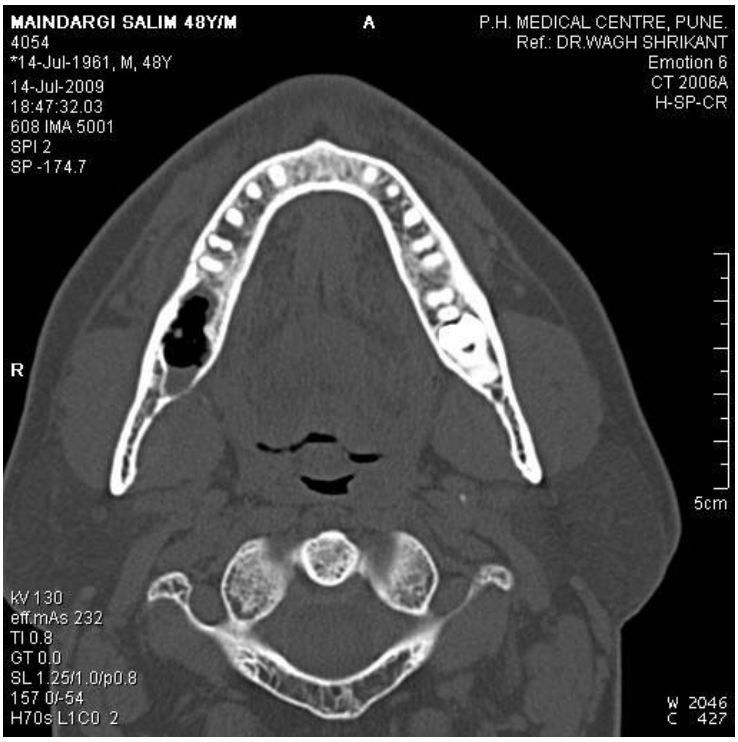

Fig. 4 Osteonecrosis of Jaw

5] Multicentric Reticulohistiocytosis

(Courtesy: Dr. Sunil Tolat (Associate Professor), Dr Arvind Kaul, Department of Dermatology and Venerology, Sasson General Hospital, Pune)

A 50-year-old army man presented with severe debilitating polyarthritis for 6 months and multiple skin lesions associated with mild itching for 4 months. These were associated with anorexia, oral ulcerative lesions, significant weight loss and intermittent high-grade fever. Clinical examination revealed mild pallor. There was firm swelling, tenderness and varying degrees of diminution of range of movements at bilateral MCP, PIP, DIP, knee, elbow and shoulder joints along with flexion deformities at DIP joints (Fig. 5). Dermatological examination revealed discrete but grouped reddish brown,

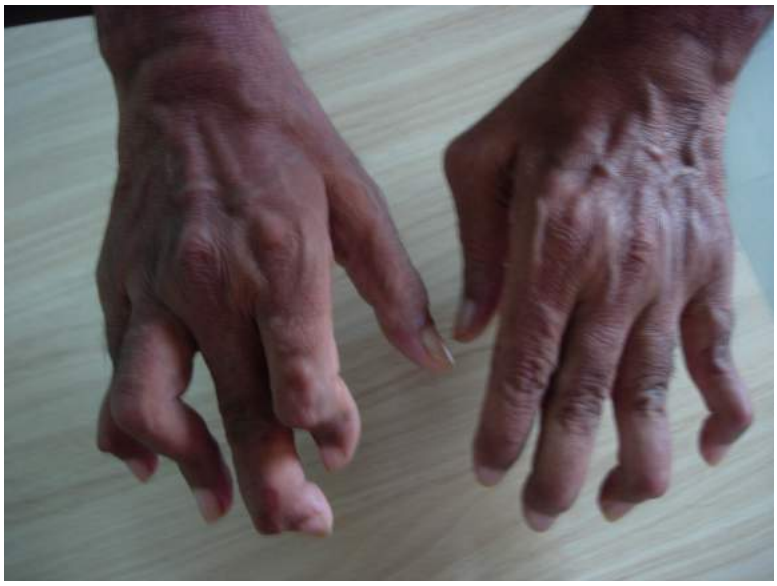

Fig.5. Hand deformities in multicentric reticulohistiocytosis

non-scaly, nontender papules and nodules of varying sizes (2mm to 2cm) over periumbilical region (Fig. 6) as well as extensor surface of forearms, back, buttocks, both knees and ankles. His hemoglobin was 8 Gm% and ESR 38 mm at 1 hour. CRP, Rheumatoid factor and other laboratory investigations were normal. X-rays of various joints revealed deformities along with soft tissue swelling. Skin biopsy showed diffuse

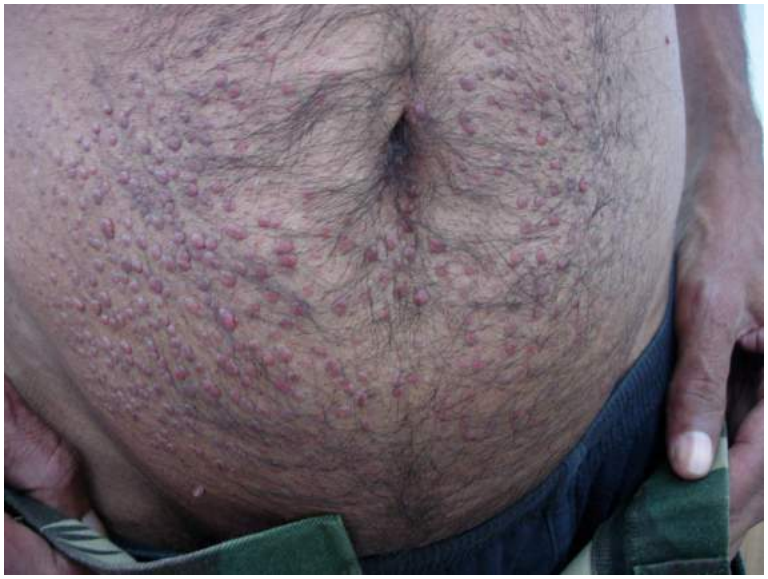

Fig 6. Skin in Multicentric Reticulohistiocytosis

infiltration of dermis by mono/multi-nucleated histiocytes with eosinophilic ground-glass cytoplasm confirming clinical diagnosis of multicentric reticulohistiocytosis.
The patient was started on monthly zoledronic acid (4 mg every intravenous) along with methylprednisolone (depot 80 mg intramuscular) every month for 6 months. He was also prescribed methotrexate 15 mg per week PO along with folic acid. Pain, swelling and skin lesions started settling down after a period of 2 months albeit with intermittent flares of skin lesions. Deformities and restriction of movements persisted at bilateral wrists, elbows, shoulders, knees and ankles as also at multiple DIP joints. Zoledronic acid was stopped at the end of 6 months whereas prednisolone (2.5 mg) and methotrexate 15 mg/week were continued. The deformities persisted despite treatment and skin rashes keep on cropping up intermittently at the end of One year.
Multicentric reticulohistiocytosis is a rare disease with destructive arthritis and skin lesions and may be associated with underlying malignancy. The disease can involve other organs too. Various drugs have been tried with limited success. The disease can remain active for many years often leaving the patient with crippling and deformed joints.

6] Risedronate induced Uveitis in patient with Paget's disease

(Courtesy: Dr. Aditya Kelkar, National Institute of Ophthalmology, Pune)

A 66-year-old male was diagnosed as a case of Paget's disease in 1999 (Fig 7). He developed stress fractures in left lower extremity twice during early 2006 (femur in January treated with closed reduction and internal fixation, and tibia in February). The patient was referred in May 2006 for rheumatology management as fracture healing was unsatisfactory. Alkaline phosphatase was 2000 IU in January 2006 and had reduced to 406 IU by May 2006. He was administered 2 intravenous injections of pamidronate 90 mg each at an interval of 2 weeks. Fractures then healed over a period of time. Patient was prescribed Risedronate 35 mg weekly later on and was continuously consuming the

same till further visit in November 2008. Patient had undergone cataract surgery with lens implantation in 1996 (right eye) and January 2007 (left eye).

Patient complained of diminished vision in both eyes in July 2008, was treated at another hospital with subconjunctival steroid injection and laser therapy. Vision improved a little but deteriorated again within a month or so. On his next visit in November 2008, he was advised to discontinue Risedronate and was referred to another ophthalmologist for evaluation.

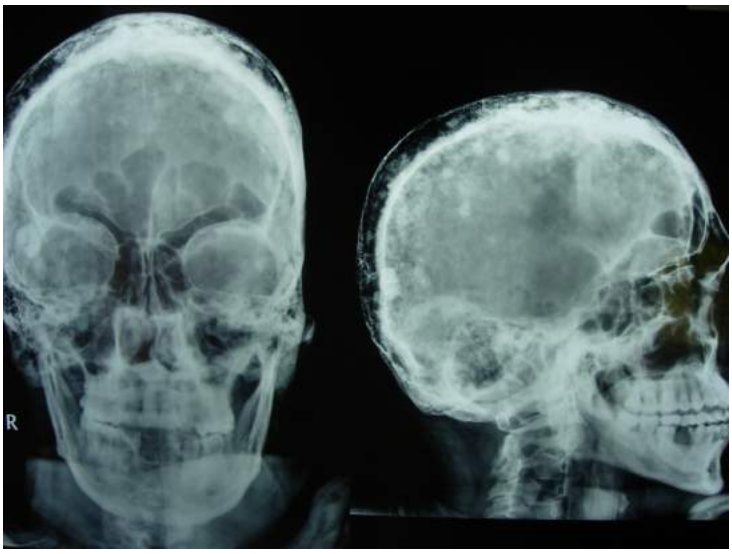

Fig 7. Paget's Disease – Skull

(diploic space, ill-defined sclerotic and lucent areas, thickened and irregular cortex)

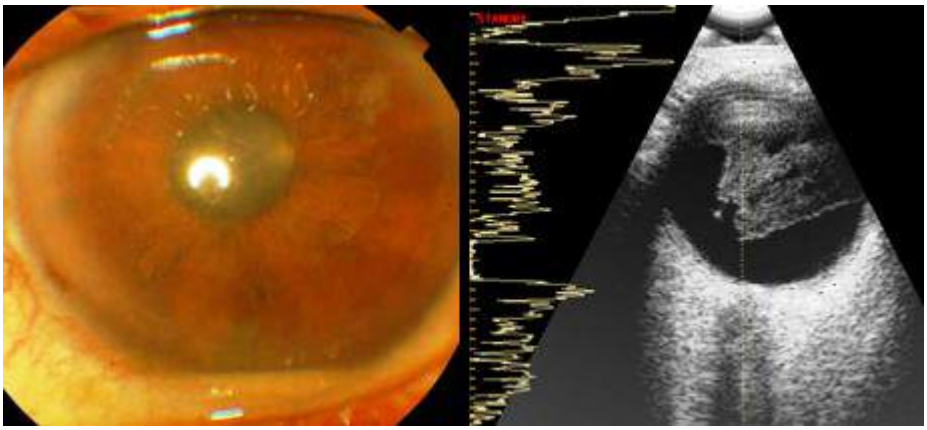

Fig. 8. Hypopyon (Left), B scan showing vitreous haze and attached retina (Right)

Ophthalmic examination revealed bilateral posterior chamber lens implants with mild iritis and vitreous haze suggestive of uveitis. Intraocular pressure was normal. He was prescribed topical steroids, anti-inflammatory and cycloplegic drugs. Anti-glaucoma drugs were also prescribed to prevent rise in intraocular pressure. His vision fluctuated over next 3 months according to severity of Uveitis. His vision returned back to normal (aided 6/9 each eye) with normal intraocular pressure in April 2009.

High nitrogen content of amino-bisphosphonates is supposed to stimulate ocular inflammation. Though rare, ophthalmic complications such as nonspecific conjunctivitis, episcleritis, scleritis, uveitis (anterior and posterior) and rejection of corneal graft have been reported. Rechallenge with bisphosphonate can lead to recurrence of symptoms.

In Rheumatology Principles and Practice (Ed. Syngle A., Deodhar A.D.), First Edition, 2010, Jaypee Brothers Medical Publishers, New Delhi