A PAINFUL HIP

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16.3.19

MRS CP 68 F

- Presented in July 2018 with a painful and swollen left lower limb, and difficulty weight bearing. No history of trauma.
- Past Medical History:
  - Osteomyelitis L) great toe 2012 (Pseudomonas)
  - Osteoarthritis
  - Venous leg ulcers
  - Iron deficiency anaemia
  - Smoker
- SHx: Lives with husband and daughter, usually mobile with a frame

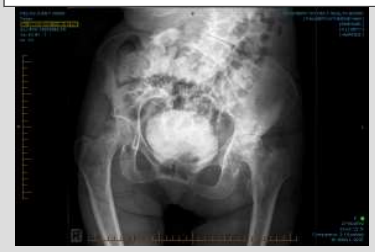
JULY 2018

- Admitted July 2018 with L) Hip pain and difficulty mobilizing
  - L) acetabular insufficiency fracture
  - Complex L) Iliac fossa mass of undetermined aetiology
  - Aspirate/Biopsy/Extensive imaging negative for infection/malignancy/no bi-refringent crystals
  - Acid fast stain and TB culture also negative
- Progress: Fracture managed non-operatively, and managed partial weight bearing and mobilized with walking aides. Lost to follow up for months.

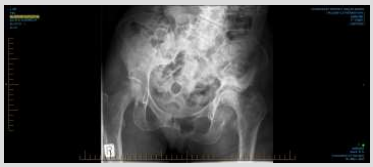
DEC 2018

- Readmitted under Orthopaedics with limited mobility and threatened independence
- O/E:
  - Unable to mobilize
  - Irritable L) Hip, minimal movement, associated soft tissue swelling L) Upper thigh
  - Incidentally, marked limited in ROM shoulders
- Rheumatology advice sought at this point

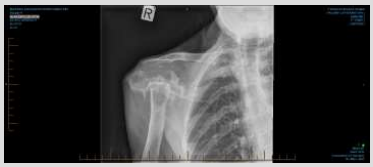
X-RAY JULY 2018



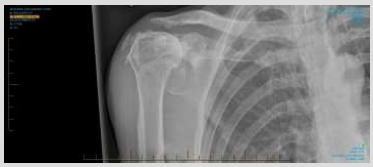
REPEAT XRAY IN DECEMBER 2018



R) SHOULDER

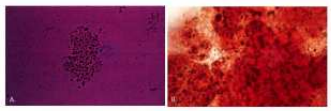


R) SHOULDER 2012



FURTHER INVESTIGATIONS

- R) Shoulder aspirate Dec 2018 – Apatite crystals seen on Alizarin red stain
- Unfortunately L) Hip aspirate was never sent



- Diagnosis:
  - Destructive arthropathy left hip with secondary left iliac fossa mass
    - Acetabular insufficiency fracture July 2018, no evidence for infection or malignancy on repeat aspirate and synovial biopsy
    - Likely basic calcium phosphate arthropathy
  - Destructive arthropathy bilateral shoulders, consistent with Milwaukee shoulder – hydroxyapatite crystals isolated on specific Alizarin stain
  - Generalised OA
  - Degenerative thoracolumbar scoliosis

BASIC CALCIUM PHOSPHATE (BCP) CRYSTAL ARTHRITIS

- Two BCP associated musculoskeletal syndromes
  - Arthritis
  - Calcific periarthritis
- OA most common arthritis associated with BCP
- Milwaukee shoulder syndrome characteristic entity

BCP CRYSTALS

- Predominant mineral type is carbonated hydroxyapatite
- BCP refers to trio of sub-micron-sized calcium phosphate crystals
  - partially carbonate-substituted hydroxyapatite
  - octacalcium phosphate
  - tricalcium phosphate
- BCP crystals are similar to normal mineral in bones and teeth and pathologic mineral in atherosclerotic plaques and calcinosis cutis

BCP ASSOCIATED ARTHRITIS

- IA crystals recognized since 1970s, initially in OA
- Milwaukee shoulder syndrome (MSS) – unique subset of patients
  - present with noninflammatory, shoulder predominant OA-like arthritis associated with BCP crystals
  - case reports in patients with autoimmune inflammatory conditions and ESRF
- By far most common entity is BCP-associated calcific periarthritis eg. calcific tendinopathy of shoulder

PATHOGENESIS

- Poorly understood
- BCP mineral inflammation occurs at sites of local tissue damage associated with injury or inflammation

RISK FACTORS

- Severe radiographic damage, CPPD crystals in pre-existing OA
- history of shoulder trauma or heavy use,
- increasing age

CLINICAL PRESENTATION

- BCP crystals are common in advanced large joint OA
  - 50% synovial fluid
  - 100% joint replacement histology
- MSS (first described 1984)
  - chronic shoulder arthritis, (may involve knees, occasionally hips)
  - large non-inflammatory shoulder effusions
  - Xray – severe joint destruction, cartilage loss and extensive rotator cuff damage
- Other conditions
  - scleroderma, CPPD arthropathy, ESRD (? overlap with calciphylaxis)
- Spectrum – OA----------→MSS

CRYSTAL IDENTIFICATION

- Synovial fluid non-inflammatory
  - BCP crystals not visible on plain polarising microscopy
- Alizarin red S staining
  - can identify Ca-containing crystals in SF
  - false positive with CPP crystals
- Other techniques – research
  - spectroscopy, transmission electron microscopy

DIFFERENTIAL DIAGNOSIS

- CPPD and gout
- Haemochromatosis
- Acromegaly
- Tenosynovial giant tumour (PVNS of shoulder)
- Neuropathic joint disease
- Rotator cuff arthropathy
- Amyloid arthropathy
- Stickler syndrome – congenital
  - Cleft palate, micrognathia, midface hypoplasia, retinal detachment, hearing loss, late-onset spondyloarthropathy

MANAGEMENT

- Limited ….as for OA
- Analgesia, NSAIDs
- Physiotherapy
- IA corticosteroid
- …. tidal lavage, colchicine
- Joint replacement

- Prognosis
  - poor, may achieve adequate analgesia but functional limitation persists