Ankylosing Spondylitis

Marie-Strümpell Disease

Bechterew's Disease

Definition

- Inflammatory disorder of unknown cause that primarily affects the axial skeleton (Sacroiliac joint, spine)
- Peripheral joints and extra-articular structures may also be involved
- Rheumatoid factor absent

Epidemiology

- Age: 2nd or 3rd decade.
- M:F ratio = 3:1

Etiology

- Idiopathic
- Immune mediated.
- Genetic factors (HLA-B27 present in > 90% cases)
- Follows exposure to bowel or urinary tract infections in predisposed people.

Pathology

- **Enthesitis**(inflammation atthe site of ligamentous and tendinous attachment to bone), is the primary site of pathology. It is often characterized by erosive lesionsthat are gradually replaced by fibro-cartilage regeneration and then finally by ossification.
- Ultimately, the joint may be totally obliterated.

Presentation

- Axial arthritis (e.g. Sacroiliitis, Spondylitis)
- Inflammatory Low back Pain > 3 months:Age of onset < 40, insidious onset, worse in the morning, worsen with prolonged inactivity or lying down, decreases with exercise, accompanied with constitutional Symptoms (Anorexia, malaise, Low grade fever)
- Morning sti ffness> 30 minutesin scroiliac joints and spine, improve with exercise
- Arthritis of proximal joints especially hip and shoulder
- Enthesitis of tendons and ligaments attached to bones, mainly in spine, sometimes back of the heel
- Spondylodiscitis
- Arthritis of costo-chondral junction

Complications

- Loss of motion and deformity as life progresses (thoracic kyphosis, forward stoop of neck or flexion contractures at the hips, compensated by flexion at the knees)
- Osteoporosis
- Spinal fracture with even minor trauma to the rigid, osteoporotic spine.

Tests and measurements for AS

1. Cervical mobility
   a. Occiput-to-wall distance(normal:0)
   b. Cervicalrotation (normal : 70-90°)
2. Thoracic mobility
   a. Chest expansion**:** normal ≥ 5cm at 4th intercostal space
3. Lumber mobility
   a. Modified Schober index**:** Measure the distracti on of 2 marks5cm below and 10cm above the dimples of Venus. when the patient bends forward as far as possible, keeping the knees straight (normal ≥ 5cm)
   b. Finger-to-floor distance: (normal ≤ 15cm)
   c. Lumbar lateral flexion: (normal ≥ 10cm)
4. For sacroiliitis
   a. Pelvic compression test: Bimanual compression of iliac crests towards midline →SI pain
   b. Faber test : flexion, abduction, external rotation of hip with heel on

contralateral knee → contralateral SI pain

c. Gaenslen Test:Flexion of hip and knee on one side and extension of hip on another side→ ipsilateral pain

Extra-skeletal Manifestation

- Acute anterior uveitis (Most common): typically unilateral, causing pain, photophobia, and increased lacrimation.
- Cataracts and secondary glaucoma
- Colitis or ileitis.
- Aortitis, Aortic insufficiency.
- Third-degree heart block.
- Subclinical pulmonary lesions.
- Slowly progressive upper pulmonary lobe fibrosis.
- Retroperitoneal fibrosis.
- Prostatitis.

Diagnosis

Modified New York Criteria (1984): criterion 4 + any of 1/2/3

1. Inflammatory low back pain > 3 months

2. Limited moti on of lumbar spine in sagitt al and frontal planes

3. Limited chest expansion (<2.5cm at 4th ICS)

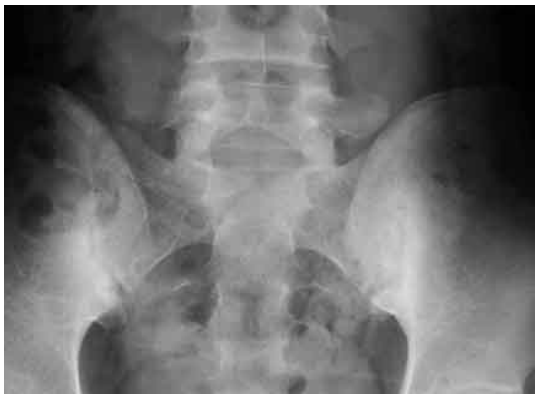
- 4. Definite radiologic sacroiliiti s

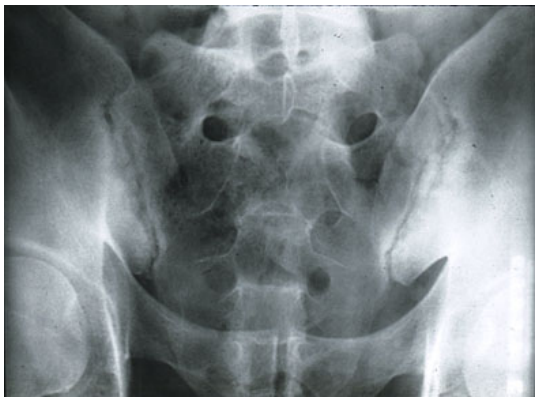
Laboratory Investigations

- HLA B27: present in ≈ 90% of pati ents.
- ESR and CRP – often elevated.
- Mild anemia.
- Elevated serum IgA levels.
- ALP and CPK raised.

Radiologic Examination

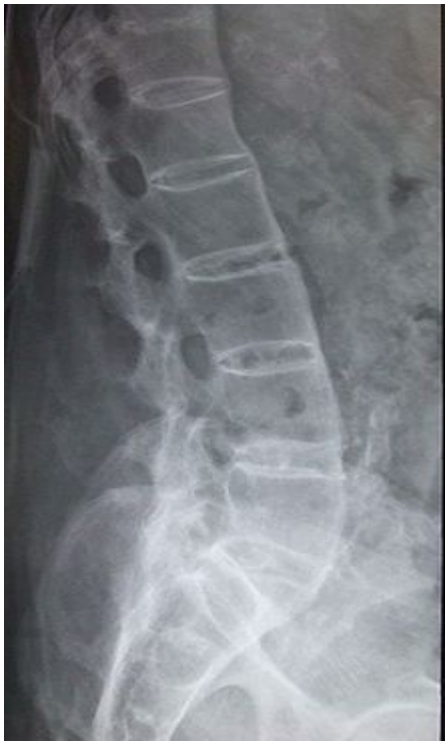
1. Sacroiliitis
    - Early:blurring of the cortical margins of the sub-chondral bone
    - Followed by erosions and sclerosis.
    - Progression of the erosions leads to “pseudo widening” of the joint space
    - As fibrous and then bony ankylosis supervene, the joints may become obliterated.
    - The changes and progression of the lesions are usually symmetric.
    - Seen in Ferguson's View (specialized sacroiliac view).
    - Dynamic MRI is the procedure of choice for establishing a diagnosis of sacroiliitis.

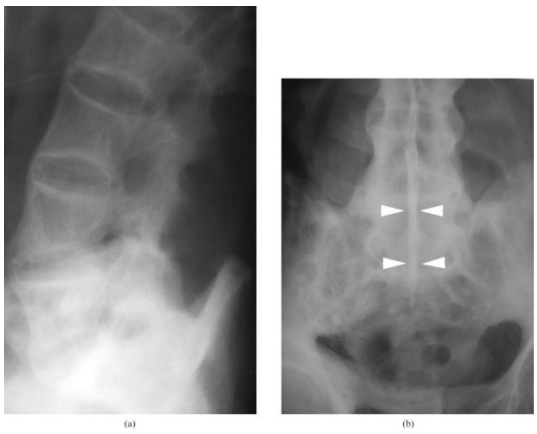




2. Lumbar spine
    - Loss of lordosis/ straightening
    - Diffuse osteoporosis
    - Reactive sclerosis- caused by osteitis of the anterior corners of the vertebral bodies with subsequent erosion (Romanus lesion), leading to “squaring” of the vertebral bodies.
    - Ossification ofsupraspinous and interspinous ligaments “dagger Sign”.

- Formation of marginal syndesmophytes
- Later Bamboo spine appearance when ankylosis of spine occurs.
- Odontoid erosion.





Treatment

1. Regular physical therapy
2. NSAIDS: Indomethacin (up to maximum of 50 mg PO tid)
3. Sulfasalazine: 2-3 g/d-Effective for axial and peripheral arthritis
4. Methotrexate: 10 - 25 mg/wk- primarily for peripheral arthritis
5. Local Corticosteroids injection for persistent synovitis and enthesopathy
6. Anti-TNF-α therapy – Infliximab (chimeric human/mouse anti-TNF-α monoclonal antibody), or Etanercept (soluble p75 TNF-α receptor–IgG fusion protein)