THE NORTHERN HOSPITAL

ANKYLOSING SPONDYLITIS

"Mariana in the Moated Grange" 1850-51, Oil on Panel, Tate gallery London. John Everett Millais, Pre-Raphaelite, (1829 -1896)

ANKYLOSING SPONDYLITIS

Introduction

Ankylosing spondylitis is one of a family of related and overlapping rheumatic diseases known as the spondyloarthropathies that include:

- Ankylosing spondylitis
- Reiter's disease
- Behcet's syndrome
- Psoriatic arthritis
- Arthritis associated with inflammatory bowel disease

This group of diseases shares various characteristics including:

- An association with HLA-B27
- The presence of enthesitis as the basic pathologic lesion, (see below)
- A familial aggregation.
- **Negative** rheumatoid factor tests.
- There is overlap between the different conditions, i.e. it is likely that more than one of these conditions may be present in the same family or in the same patient.

Ankylosing spondylitis is characterized by its predominance of effects on the axial skeleton and the sacroiliac joints in particular.

Ankylosing spondylitis is the *most common* of the spondyloarthropathies.

Pathophysiology

The spondyloarthropathies are chronic inflammatory diseases involving the bones,

- In particular, the sacroiliac joints and axial skeleton, and to a lesser degree, the peripheral joints.

and certain extra-articular involvement ,including

- The eyes, skin, lung, and cardiovascular system.

Cytokines, in particular tumor necrosis factor-a (TNF-a) are important in the chronic inflammatory process by leading to fibrosis and ossification especially at sites of "enthesitis".

In ankylosing spondylitis the major pathological processes that occurs is referred to as "enthesitis". This is an inflammatory reaction that that tends to occur at points of ligamentous, tendon or joint capsule attachment to bone and results in fibrosis, ossification and consequent ankylosis.

Causes

- The etiology is essentially unknown but probably involves the interaction of both genetic and environmental factors.
- They are strongly associated with human leukocyte antigen (HLA) B27 and its various genotypic subtypes.

Complications

1. Skeletal
    - Bilateral sacroiliitis and eventual ankylosis.
    - Thoracolumbar ankylosis, leading to stiffness and deformity.
    - Costo-vertebral ankylosis, leads to a reduction in chest expansion with a consequent restrictive lung impairment.
    - Peripheral joint involvement tends to be larger proximal joints, hips, shoulders rather than the smaller peripheral joints of the hand or feet.
2. Eyes
    - Anterior uveitis is the most frequent extra-articular manifestation.
3. Skin
    - There is an association with psoriasis.
4. CVS
    - Aortic incompetence secondary to aortitis of the ascending aorta.
    - Pericarditis
    - Conduction defects
5. Respiratory
    - Pulmonary fibrosis, (predominantly of the *upper* lobes).
6. GIT
    - There is an association with inflammatory bowel disease.

7. Tendons

- Inflammatory changes may occur at tendinous insertions, particularly resulting in plantar fasciitis and Achilles tendonitis and patella tendonitis at the tibial tuberosity.

Clinical Assessment

From the ED perspective the important issues will be:

1. The identification of possible early presentations of the disease and consequent early Rheumatological referral.

2. An awareness of the possible secondary or non-skeletal systemic complications in those who have diagnosed and established disease.

Possible early presentations

Symptoms typically begin in younger age groups, between 15 and 30 years.

The onset of symptoms is insidious.

1. Systemic "constitutional" symptoms:

   - Fever

   - Myalgias, arthralgias, malaise may occur, to a mild degree, (in contrast to rheumatoid arthritis where these symptoms can be significant)

2. Inflammatory back pain:

   - This is the most common symptom and the first manifestation in approximately 75% of patients.

   - Symptoms will most commonly be manifested in the **sacroiliac joints** or the **lower lumbar spine.**

   - Symptoms include **morning stiffness lasting at least 30 minutes**, and diffuse nonspecific radiation of pain into both buttocks.

   - Symptoms are not improved with rest and in fact improvement of symptoms may occur with moderate physical activity, (in distinction to "mechanical" back problems)

   - There may be loss of normal lumbar lordosis, due to muscle spasm.

   - Symptom onset is insidious, (in distinction to mechanical back pain which tends to be far more acute)

- There will be limitation of spinal movement in all three planes.

  Schober's test

  Although Schober's test is nonspecific, it is useful for measuring spinal mobility.

  The test is performed by marking the patient's back over the L5 spinous process (between the posterior superior iliac spines) and 10 cm above this point. The patient then is asked to bend forward.

  The distance between the two marks should increase by 5 cm or more in normal persons. An increase of less than 5 cm suggests decreased range of motion of the lumbar spine.

- Most patients have mild chronic disease or intermittent flares with periods of remission. The spinal disease rarely is active persistently.

3. Loss of chest expansion (<5 cm at the level of the 4^{th} intercostal space) usually is found only in patients with late-stage disease and, generally, is not helpful in making an early diagnosis.

Investigations

The need for investigation in the ED will depend on the index of suspicion for the condition or the suspicion of the presence of systemic complications.

Although diagnostic criteria for the spondyloarthropathies have been developed for research purposes, the criteria rarely are used in clinical practice.

Diagnosis is based primarily on the history and physical examination.

There are no *specific* diagnostic tests for spondyloarthropathies.

Evidence of sacroiliitis or spondylitis may be seen on radiographs of the pelvis and lumbar spine.

Blood tests:

1. FBE

   - There may be an associated anemia of chronic disease.

2. U&Es/ glucose

   - Establish baseline renal function.

3. Inflammatory markers.

   - ESR may be elevated.

- CRP may be elevated.

4. Immunological tests:

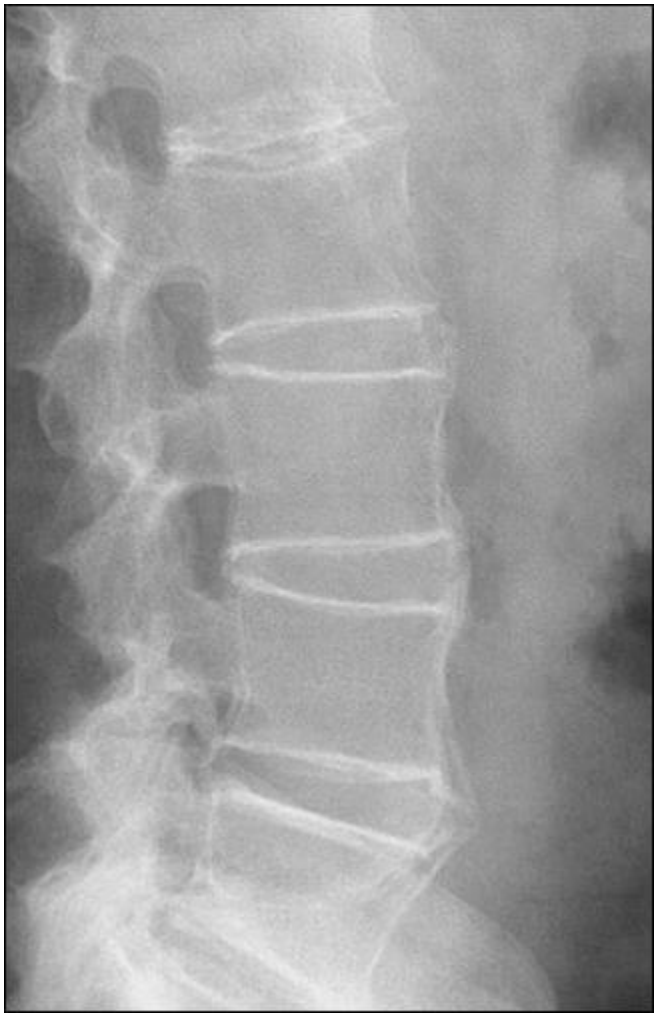
   - HLA B-27 testing provides supportive evidence, (but does not make the diagnosis)

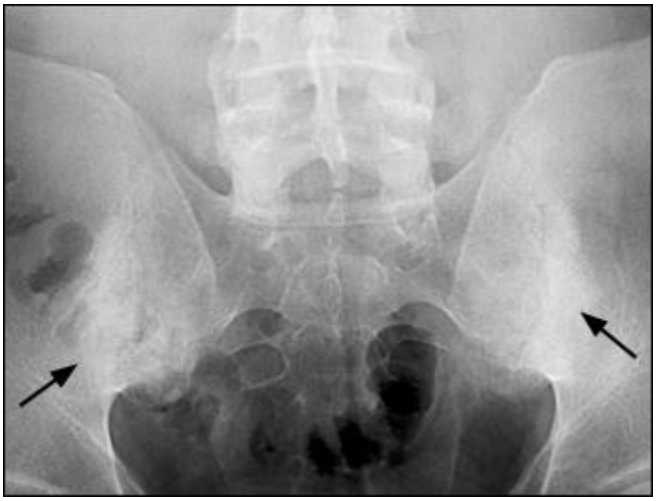
   - Rheumatoid factor is **not** elevated.

Joint aspiration:

- The synovial fluid typically is inflammatory (more than 2,000 white blood cells per mL, with a predominance of neutrophils), but this finding is nonspecific.

Radiology:





Above, sacroiliitis, with loss of joint space and adjacent sclerosis.

Left, Ossification of the annulus fibrosis, showing the typical "bamboo" spine appearance.

Radiographic features of ankylosing spondylitis include

1. Bilateral symmetric sacroiliitis

- There is initial sclerosis progressing to erosive changes and finally to total ankylosis or fusion of the sacroiliac joints

2. Vertebral column, lumber spine in particular.

   - Inflammation at the site of insertion of the annulus fibrosus can result in osteitis of the anterior vertebral margins, giving a so-called "shining corners" sign.

   - Later on syndesmophytes may form, bony bridges between the vertebral bodies within the thoracolumbar regions, resulting in the so-called "bamboo" spine appearance.

Magnetic resonance imaging and computed tomography scan

These are not part of the "routine" evaluation of patients with ankylosing spondylitis, however:

- MRI or CT scan of the sacroiliac and peripheral joints may reveal evidence of *early* sacroiliitis, erosions and enthesitis that are not apparent on standard radiographs.

- Due to the insensitivity of standard radiographs in the clinical setting of acute back pain in *advanced* AS, an MRI or CT scan may be useful in making the diagnosis of a spinal fracture in patients with *late-stage* spinal disease.

  The threshold for these imaging modalities should be lower in cases where the cervical spine needs to be cleared following trauma.

Management

1. Physiotherapy

   - This will be essential to help prevent longer term disability.

2. Drug options

   For full prescribing details see Rheumatology Therapeutic Guidelines.

   NSAIDS

   - These are a good first line option, especially in undiagnosed cases.

   Sulfasalazine

   - This is considered second line agent in those who do not respond to NSAIDs or are unable to take them.

Methotrexate

- Although not well studied, methotrexate may be beneficial in patients with prominent peripheral arthritis.

Steroids

- Oral corticosteroids in conventional dosages are of little value in the treatment of ankylosing spondylitis, but intra-articular corticosteroid injections may provide rapid and sustained relief in isolated inflamed joints.

Tumor necrosis factor-alpha inhibitors.

- These are monoclonal antibody preparations. [2]

- The DMARD agents used in rheumatoid arthritis have not proved very useful in ankylosing spondylitis with respect to modifying disease progress. The newer tumor necrosis factor-alpha inhibitors however do show some early promise in this regard.

References

1. Kataria R.K. Spondyloarthropathies. American Family Physician, June 15, 2004.

2. Schachna L, editorial, The anti-TNF revolution in ankylosing spondylitis. MJA 2004; 181 (10): 529-530

3. eMedicine Website, Rheumatology, December 2005.

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