CASE TWO

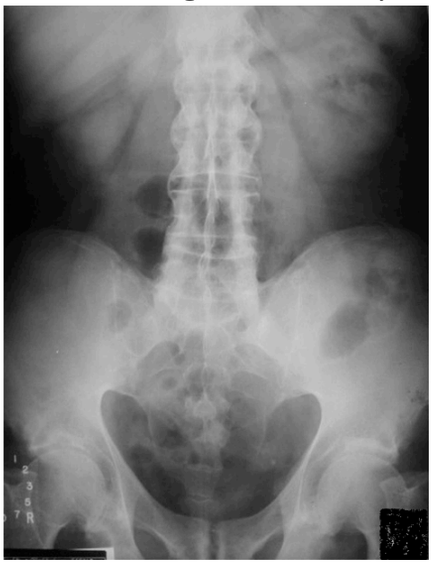
Short case number: 3_26_2
Category: Musculoskeletal System & Skin
Discipline: Medicine
Setting: General Practice
Topic: Seronegative Spondyloarthropathies.

Case

Dave Higgins is a 21 year old keen hockey player has just returned from the interuniversity games. He was unable to play in the finals because of back pain. He explains that he has had the pain for a few months. It is in his lower back, he comments that before it would improve once he started playing, but over the last few days he is just too stiff and sore. There is no history of trauma and his past medical history is unremarkable.

Questions

1. The degree of pain and stiffness in Dave's back and his young age, prompt you to consider ankylosing spondylitis. What are the key features of history and examination that would support this diagnosis? What other forms of arthritis would you consider in your assessment?
2. Why are these types of arthritis termed seronegative? What further investigations would you undertake to confirm the diagnosis?



3. Identify the features of ankylosing spondylitis seen in the radiograph of the lumbar spine.
4. You explain to Dave, that he has ankylosing spondylitis; he asks you what it is and why he has pain in his back and his chest. What would you explain to him?
5. Describe the non-articular problems that can occur with seronegative arthropathies.
6. Dave is not keen on medications and would like to avoid them if he could, outline a management plan for Dave that outlines the key components of his management and explains when medications would be used and why.

Suggested reading:

- Kumar P, Clark ML, editors. Kumar & Clark's Clinical Medicine. 8th edition. Edinburgh: Saunders Elsevier; 2012.
- Colledge NR, Walker BR, Ralston SH, Penman ID, editors. Davidson's Principles and Practice of Medicine. 22nd edition. Edinburgh: Churchill Livingstone; 2014.

ANSWERS

1. The degree of pain and stiffness in Dave's back and his young age, prompt you to consider ankylosing spondylitis. What are the key features of history and examination that would support this diagnosis?

   The onset is usually insidious, over months or years, with recurring episodes of low back pain and marked stiffness. Radiation to the buttocks or posterior thighs may be misdiagnosed as sciatica. Unlike mechanical back pain, symptoms extend over many segments and are axial and symmetrical in distribution. Symptoms are most marked in the early morning and after inactivity and are relieved by movement. Although the lumbosacral area is usually the first and worst affected region, some patients present with mainly thoracic or neck symptoms. The disease tends to ascend the spine slowly and eventually, after several years, the whole spine may be affected. As the spine becomes progressively ankylosed, spinal rigidity and secondary osteoporosis predispose to spinal fracture, presenting as acute, severe, well-localised pain. Secondary spinal cord compression is a rare complication.
   Early physical signs include failure to obliterate the lumbar lordosis on forward flexion, pain on sacroiliac compression, and restriction of movements of the lumbar spine in all directions. As the disease progresses, stiffness increases throughout the spine, and chest expansion frequently becomes restricted. Spinal fusion varies in its extent but in a few patients results in marked kyphosis of the dorsal and cervical spine that can interfere with forward vision. This may prove incapacitating, especially when associated with fixed flexion contractures of hips or knees.

2. What other forms of arthritis would you consider in your assessment?

   - Reactive arthritis, including Reiter's syndrome
   - Psoriatic arthropathy
   - Arthritis associated with inflammatory bowel disease (Crohn's disease, ulcerative colitis)

3. Why are these types of arthritis termed seronegative? What further investigations would you undertake to confirm the diagnosis?

   This term is applied to a group of inflammatory joint diseases listed above that are distinct from RA. These diseases are thought to share a similar pathogenesis as they show considerable overlap and similarity of articular and extra-articular clinical features and a striking genetic association with the histocompatibility antigen HLA-B27.
   Diagnostic features include:
   - ESR and CRP are usually raised.
   - Serum rheumatoid factor is negative or present in low titre.
   - Radiographic signs provide the strongest investigational evidence but may take years to develop. Sacroiliitis is often the first abnormality, beginning in the lower synovial parts of the joints with irregularity and loss of cortical margins, widening of the joint space and subsequently sclerosis, narrowing and fusion. Lateral thoracolumbar spine X-rays may show anterior 'squaring' of vertebrae due to erosion and sclerosis of the anterior corners and periostitis of the waist. Bridging syndesmophytes are fine and symmetrical and follow the outermost fibres of the annulus. Ossification of the anterior longitudinal ligament and facet joint fusion may also be visible. The combination of all these features results in the typical 'bamboo' spine. Erosive changes may be seen in the symphysis pubis, the ischial tuberosities and peripheral joints. Osteoporosis and atlantoaxial dislocation can occur.

4. Identify the features of ankylosing spondylitis seen in the radiograph of the lumbar spine.
   Bamboo' spine of severe late ankylosing spondylitis. Note the symmetrical marginal syndesmophytes, sacroiliac joint fusion and generalised osteopenia.

5. You explain to Dave, that he has ankylosing spondylitis; he asks you what it is and why he has pain in his back and his chest. What would you explain to him?

   Ankylosing spondylitis is a chronic, multi-system inflammatory disorder of the sacroiliac (SI) joints and the axial skeleton. It is called a seronegative spondyloarthropathy and sometimes found in association with other problems including reactive arthritis, psoriasis, juvenile chronic arthritis, ulcerative colitis, and Crohn disease.
   The aetiology of this condition is not understood completely but there does seem to be a strong genetic predisposition. A direct relationship between AS and the major histocompatibility human leukocyte antigen (HLA)-B27 has been identified.

6. Describe the non-articular problems that can occur with seronegative arthropathies.

   - Anterior uveitis (25%) and conjunctivitis (20%)
   - Prostatitis (80% men)-usually asymptomatic
   - Cardiovascular disease
     - Aortic incompetence
     - Mitral incompetence
     - Cardiac conduction defects
     - Pericarditis
   - Amyloidosis
   - Atypical upper lobe pulmonary fibrosis

7. Dave is not keen on medications and would like to avoid them if he could, outline a management plan for Dave that outlines the key components of his management and explains when medications would be used and why.

   The aims are to relieve pain and stiffness, maintain a maximal range of skeletal mobility and avoid deformity. Education and appropriate physical activity are the cornerstones of management. Early in the disease patients should be taught to perform regular daily back extension exercises, including a morning 'warm-up' routine, and to punctuate prolonged periods of inactivity (e.g. driving, computer work) with regular breaks. Swimming is ideal exercise. Poor bed and chair posture must be avoided.

   NSAIDs are effective in relieving symptoms but do not alter the course of the disease. A long-acting NSAID at night is particularly helpful for marked morning stiffness. The slow-acting antirheumatic drugs sulfasalazine, methotrexate or azathioprine may control persistent peripheral joint synovitis but appear to have little or no impact in suppressing axial disease. However, recent studies have shown that anti-TNF therapy may improve the symptoms and signs of ankylosing spondylitis, including spinal mobility.

   Local corticosteroid injections can be useful for persistent plantar fasciitis and other enthesopathies. Oral steroid may occasionally be required for acute uveitis but should otherwise be avoided. Severe hip, knee or shoulder restriction may require surgery. Total hip arthroplasty has largely obviated the need for difficult spinal surgery in those with advanced deformity.