Case Report

Ankylosing spondylitis: A Case Report with Review of Literature

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ABSTRACT

Background: Ankylosing spondylitis is a relatively common worldwide chronic inflammatory disease that usually affects spine, sacroiliac joints in addition to peripheral joints and affects multiple other tissues, which can progress to bony fusion of the spine.

Objective: To fill the gap of the absence of a published description of this condition in Libyan patients and to compare of the clinical presentation and other aspect of the condition in a Libyan patient with what is known worldwide.

Materials and Methods: A case report of a 60 year old Libyan male who has Ankylosing spondylitis and then review the available literature.

Results: The patient has been having a chronic low back pain and stiffness for the last 26 years. His symptoms are most severe in morning and improve with movement. He developed stiffness and limitation of movement of the neck and restriction of movements of the shoulder and hips. Examination showed kyphosis and limited neck movement and decreased chest expansion. His score on Bath Ankylosing Spondylitis Disease Activity Index is 6. His x-rays showed bamboo spine appearance of spine and osteoarthritis in both shoulder joints and hip joints. His presentation fits both European Spondyloarthropathy Study Group diagnostic criteria for spondyloarthropathies and the modified New York Criteria.

Conclusion: Our Libyan patient shows a classical clinical and radiological picture of the condition and the diagnostic criteria and severity index could be applied easily to his cases.

Key words: Ankylosing spondylitis, Spondyloarthritis, back pain, back stiffness, bamboo spine.

Ankylosing spondylitis is one of the commonest spondyloarthritis with prevalence that range between 0.5% and 0.9% worldwide[1,2,3].The prevalence differs from one region to another. In china the prevalence of Ankylosing spondylitis is about 0.24% and in Europe it can reach up to 1.4%.[4]The most common regions affected by Ankylosing spondylitis are the spine and the sacroiliac joints. In addition to those

areas, Ankylosing spondylitis can affect the tendons and ligaments, peripheral joints, eyes, skins and bowel causing enthesitis, arthritis, anterior uveitis, prostatitis, psoriasis and inflammatory bowel diseases[1].

There is a strong role of genetic inheritance in the development of Ankylosing spondylitis and twin studies have shown that the heritability of Ankylosing spondylitis can reach up to more than 90%[1,5]. Studies have linked human leukocyte antigen (HLA)-B27 with Ankylosing spondylitis[1,5,6]. HLA-B27 positive patients represent more than 90% of patients with Ankylosing spondylitis[7] and they usually have more severe clinical manifestations when compared to HLA-B27 negative patients[6]. HLA-B60 and

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HLA-DR1 were also found to be associated with Ankylosing spondylitis. An infectious etiology is thought to play a role in the development of Ankylosing spondylitis and the suspected pathogens are Klebsiellapneumoniae and Escherichia coli[3].The pathogenesis of Ankylosing spondylitis is not yet completely understood[7,8]. Several hypothesis are present like arthritogenic peptide theory, aberrant folding of the heavy chain of HLA-B27, autodisplay hypothesis and β2m deposition hypothesis but none of them is final yet[7].

CASE PRESENTATION:
Our patient is a 60 year old Libyan male who reported to Sebha Medical Center with complaints of chronic fatigue for the last 10 years and shortness of breath of 4 days duration. His shortness of breath has been exacerbated on lying flat and has been associated with swelling of lower limbs. There is no history of cough, expectoration, wheezing, haemoptysis, and chest pain, pain in feet, heel, or bony prominences on the side of hips or above the pelvis on the sides. He is a known case of Diabetes mellitus and has been on insulin. He has been having chronic low back painand stiffness with limitations of spine movements for the last twenty six years. There is no history of radiation of pain to buttock or behind thigh. The symptoms have been most marked in early morning after sleep and used to get relieved by movements earlier but now movements of spine have become very restricted in all directions. There is no weakness of limbs. Later he developed pain and stiffness in neck and has gradually worsened to inability to move the neck in all the directions. He cannot lift his arms above the shoulders because of painful restriction. He cannot sit in squatting position because of painful restriction at hip joint. There is no history of pain or redness of eyes or urinary complaints. There is no history of bowel complaints or skin lesions.

Table 1: Bath Ankylosing Spondylitis Disease Activity Index

Question	Score	Actual score in the Patient
1. How would you describe the overall level of fatigue or tiredness you have experienced?	1–10	9
2. How would you describe the overall level of neck, back or hip pain you have had?	1–10	9
3. How would you describe the overall level of pain and swelling you have had in joints other than the neck, back or hip?	1–10	9
4. How would you describe the overall level of discomfort you have had from any areas tender to touch or pressure?	1–10	9
5. How would you describe the overall level of discomfort you have had from the time you wake up?	1–10	9
6. How long does your morning stiffness last from the time you wake up?	0 hr to2+ hrs	9
The patient is asked to complete each question. The score is calculated by taking the average of all 6 questions, where duration of morning stiffness in minutes is coded in 12-minute increments from none (1) to 120 (10). Online calculators are also available at http://basdai.com		

He had gone abroad and twenty years ago he had received physiotherapy for one and a half year, but his problem continued to worsen with time. Table 1 shows the questions of Bath Ankylosing Spondylitis Disease Activity Index and the actual score that the patient had according to his answers on those questions.

On examination: Conscious, oriented and afebrile. General condition: Stable, blood pressure 150/80 mm Hg, bilateral pitting oedema of lower limbs without tenderness or erythema or raised local temperature. CVS examination revealed muffled heart sounds but no murmurs or extra sounds. Examinations of his chest, abdomen and central nervous system were normal apart from bilateral decreased chest expansion (chest expansion-1.5 cm). Examination of his musculoskeletal system revealed kyphosis of thoracic spine and severe limitation of cervical spine movements (stiff neck). He could not touch the bed with his head because of marked stiffness of cervical spine (forward stoop of the neck) (occiput-to-wall distance-17cm). Movements of both shoulder and hip joints were painful and restricted. He could not move his lumbar spine i.e. could not bend forward, backward or sideways (anteriorly, posteriorly, and laterally). There was tenderness over both sacroiliac joints, spinous processes, iliac crests and greater trochanters. He could not sit i.e. he could not flex his hip joints and any attempt at flexion of hip joints elicited severe pain.

The modified Schober test revealed no increase in distance. [The modified Schober test is a useful measure of lumbar spine flexion. The patient stands erect with heels together, and marks are made on the spine at the lumbosacral junction (identified by a horizontal line between the posterosuperior iliac spines) and 10 cm above. The patient then bends forward maximally with knees fully extended, and the distance between the two marks is measured. This distance increases by 5 cm in the case of normal mobility and by <4 cm in the case of decreased mobility.]

His complete blood count was normal with WBC = 9,000/cmm, RBC=3,500,000/cmm, Hb-13Gm%, platelets =220,000/cmm. Random blood sugar= 531mg/dl, fasting blood sugar= 209 mg/dl, HbA1C= 8.0%, Blood Urea=29mg/dl, and serum creatinine=0.76mg/dl. Liver functions tests showed low total protein (5.0 Gm%) and low albumin (2.5Gm%). Total serum bilirubin- 0.7mg/dl, unconjugated bilirubin-0.5mg/dl and conjugated bilirubin- 0.2mg/dl, AST- 26U/L, ALT-29U/L and ALP- 91U/L. His serum cholesterol was mildly elevated (224mg/dl) but serum triglyceride was within the normal range (149mg/dl). ESR was high (39mm/1hr), but tests for RF, ANA and CRP were negative. His general urine examination showed large number of pus cells (20-40/HPF) and RBC (over 50/HPF). His ECG was normal.

Box 1: European Spondyloarthropathy Study Group diagnostic criteria for spondyloarthropathies

Inflammatory spinal pain
OR
Synovitis asymmetrical or predominantly in the lower limbs)
AND
One or more of the following:
Positive family history
Psoriasis
Inflammatory bowel disease
Alternate buttock pain
Enthesopathy
Sacroiliitis

A chest X-ray was taken and it showed abnormal thoracic spine (bamboo spine) (Figure 1). The patient's cervical spine, both shoulder joints and pelvis were imaged using X-rays and all the images revealed multiple abnormalities (Figure 2 and 3). Echocardiography revealed mild

dilatation in the patient's right atrium and right ventricle with mild tricuspid regurgitation with evidence of mild pulmonary artery hypertension. No abnormalities were found on ultrasonography examination of the patient's abdomen and pelvis.

DISCUSSION:
Ankylosing spondylitis, which is also known as Bechterew (Bekhterev's) disease and as Marie Strumpell disease[9], has been found in Egyptian mummies and that means it was present thousands of years ago, but it wasn't described in medical

Figure1: The X-ray chest of the patient showing bamboo spine appearance of the spinal column. The patient could not elevate his head for the image because of fixed flexed cervical Spine.

literature until 1559 AD[3]. Even though it was present and known for long time, its causes and mechanisms are not completely understood and that leads to it being one of the diseases that don't respond well to the currently available treatment in addition to it being difficult to diagnose[3].

Ankylosing spondylitis belongs to a family of the diseases called Spondyloarthropathies. This family includes multiple other conditions in addition to ankylosing spondylitis like reactive arthritis (ReA), psoriatic arthritis, arthritis related to inflammatory bowel diseases, as well as undifferentiated spondyloarthropathy[7].
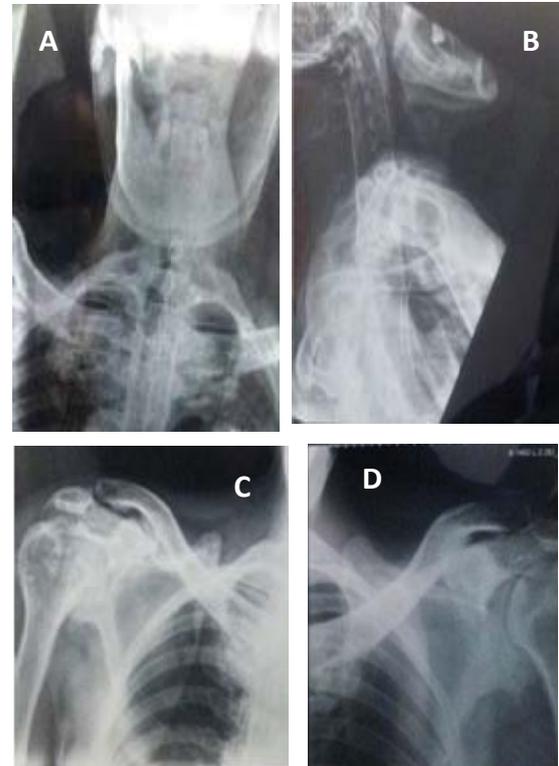

Figure2: Lateral (A) and Antero-posterior (B) X-rays of the patient's cervical spine showing bamboo spine appearance of cervical spine. The patient could not elevate his head enough for the image. AP views of right (C) and left (D) shoulders showing osteoarthritis in both shoulder joints.

Spondyloarthropathies are usually defined according to the criteria of the European Spondyloarthropathy Study Group (Box1)[10].

Ankylosing spondylitis is more common in males than in females and its first symptoms usually appears in the third decade of the patient's age[12].Most patients are diagnosed before they reach 40 years of age and the average delay since the onset of the symptoms till the time of the diagnosis is between 5 and 8 years[13,14]. Some causes that might explain that delay

Table 2: The modified New York Criteria which is used to diagnose Ankylosing Spondylitis Criteria

Clinical criteria

   a. Low back pain and stiffness for more than 3 months which improves with exercise, but is not relieved by rest.

   b. Limitation of motion of the lumbar spine in both the sagittal and frontal planes.

   c. Limitation of chest expansion relative to normal values corrected for age and sex. (<5cm=Abnormal in young adult)

Radiologic criterion of sacroiliitis grade greater than or equal to 2 bilaterally or sacroiliitis grade 3-4 unilaterally. Grades are as follows:

   a. 0 = normal

   b. 1 = suspicious changes

   c. 2 = minimum abnormality (small localized areas with erosions or sclerosis)

   d. 3 = unequivocal abnormality (moderate or advanced sacroiliitis with erosions, evidence of sclerosis, widening, narrowing or partial ankylosis)

   e. 4 = severe abnormality (total ankylosis)

Grading

   1. Definite Ankylosing spondylitis diagnosis if the radiologic criterion is associated with at least 1 clinical criterion.
   2. Probable Ankylosing spondylitis if:
      a. Three clinical criteria are present.
      b. The radiologic criterion is present without any signs or symptoms satisfying the clinical criteria. (Other causes of sacroiliitis should be considered.)

Figure 3: Antero-posterior X-ray of the patient's pelvis showing severe arthritis in both sacroiliac joints with sclerosis and complete fusion of the joints. It also shows destruction, sclerosis and fusion of both hip joints (right >left) and bamboo spine.

in diagnosis is that clinicians are usually not aware of the condition, that sacroiliitis doesn't appear in x-rays in the early stages of the disease and the difficulty in differentiating inflammatory and mechanical back pain[13]. There are multiple criteria groups that are available to differential inflammatory back pain from mechanical back pain like Calin's criteria, Berlin criteria and ASAS (Assessment of Spondylo Arthritis international Society) criteria[13].These criteria in general show that inflammatory back pain usually has an insidious onset, starts in an age below 40 years, starts at night, improves with exercise but not with rest, is associated with morning stiffness that last for more than 30 minutes[13].

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