

Ankylosing Spondylitis

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Spondylitis, in the broad sense, means arthritis or inflammation of the spine. The term is derived from the Greek words "spondylos," meaning vertebra, "-itis" for inflammation, and "ankylos," meaning bent or crooked. Ankylosing spondylitis, therefore, is a chronic inflammatory disease of the spine resulting in progressive stiffening with fusion of the various anatomical elements.

History

Ankylosing spondylitis has plagued man since antiquity. Ruffer and Reitti described the skeleton of a man living during the third dynasty (2980 to 2900 B.C.) whose spinal column, presumably in its entire length, was diseased and transformed into a solid block because of new bone formation in the longitudinal ligaments. A skeleton found in Nordpfalz dated (by tomb gifts enclosed) about 400 B.C., was reported by Arnold¹ as showing similar changes. Several skeletons of ancient Nubian bones from a Negroid tribe in Eastern Africa between Egypt and Abyssinia (6th-4th century B.C.) also illustrated similar changes. Skeletal remains in Guatemala around 500 A.D.² have also shown features of ankylosing spondylitis. However, many skeletons among Egyptian mummies described as having ankylosing spondylitis may have been mislabeled, as they were later shown by x-ray to have osteophytosis.

In 1691, Bernard Connor¹ first described the disease: "I have lately seen in France, part of a human skeleton consisting of the os ilium, the os sacrum, the five vertebrae of the loins, ten of the back, five entire ribs on the right side and three on the left... all of these bones which naturally are separate and distinct from one another were here so straightly and intimately joined, their ligaments perfectly bony and their articulations so effaced that they really made 1 uniform

continuous bone." From these observations, Connor deduced that the person "must have been immovable, that he could neither bend nor stretch himself out, rise up, nor lie down nor turn upon his side." Such a skeleton might appear as illustrated (Figures 1 and 2).*

Other early descriptions were reported by Wilks (1858), von Thaden (1863), Blezinger (1864), Bradhurst (1866), Virchow (1869), Harrison (1870), Flagg (1876) and Strumpell (1884).³ The most complete clinical description of the disease, however, is credited to von Bechterew, who in 1893 described what he thought was a new neurological disease characterized by stiffness of all or part of the spine, paresis of the muscles of the back, neck and extremities. Later in 1897, Strumpell reported a group of cases with progressive ankylosis of the spine and hip joints. In 1898, Marie described six cases characterized by an ascending type of ankylosis in which the sacroiliac joints became fused and the cervical spine was least and last involved. Marie was probably the first to correlate the clinical and anatomical features of what is now known as ankylosing spondylitis. In 1904, Frankel presented a full account of the disease and called it "spondylitis ankylopoietica." Valentinini in 1899 described roentgenograms demonstrating the typical bamboo spine.

Synonyms and Eponyms

In the early years this disease was variously called spondylitis rhizomelique, Marie-Strumpell's disease, spondylitis ossificans ligamentosa, von Bechterew's syndrome, adolescent or juvenile spondylitis, spodylarthritis ankylopoietica and spondylitis deformans.¹ This was so confusing that in 1941 the American Rheumatism Association (ARA) adopted the term rheumatoid spondylitis in an effort to unify the terminology in the American literature. As it turned out, that was also a poor choice because it led

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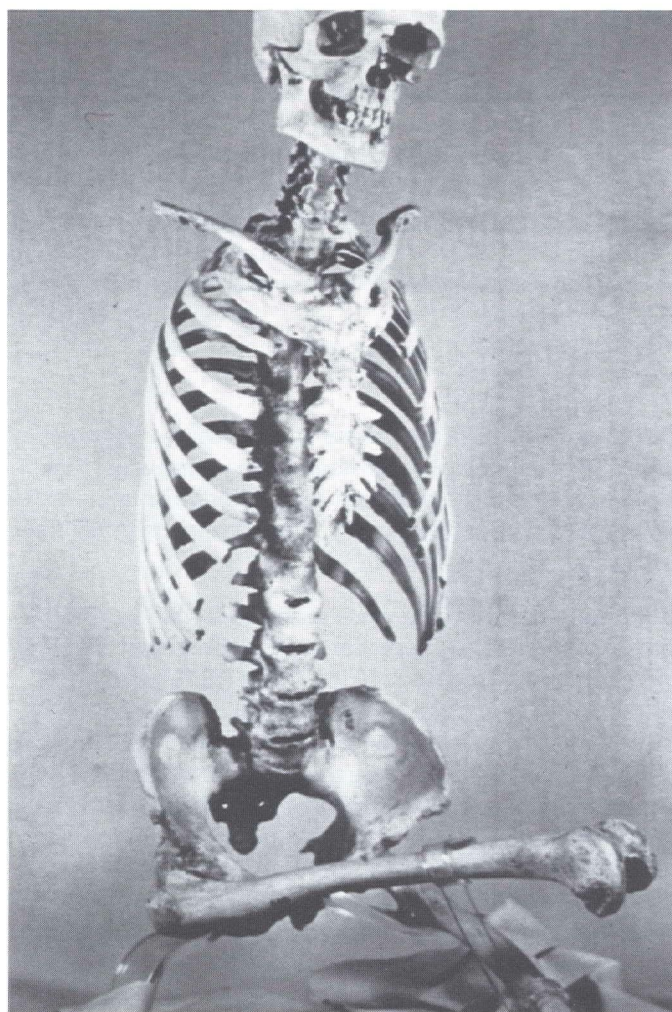


Fig. 1
Skeleton fused due to ankylosing spondylitis



Fig. 2
Enlargement of Fig. 1 showing thoracic vertebrae.

to the idea that the disease is a variant of rheumatoid arthritis. With better understanding and the recognition that it is a specific entity apart from rheumatoid arthritis, the word rheumatoid was dropped and the current name of ankylosing spondylitis was adopted.

Clinical Presentation

Ankylosing spondylitis shows a striking predilection for men. It has a relative incidence of 7:1, and its prevalence is estimated to be approximately four in 1000 Caucasian males. The onset occurs most frequently during late adolescence and early adulthood, generally before age 40. It very rarely begins after 50.

A hereditary predisposition for the disease also exists. It is inherited as a single dominant autosomal factor with 70%

penetrance in men and 10% in women, and prevalence is said to be 30 times higher among relatives of patients with spondylitis. In recent years a positive correlation has been found between ankylosing spondylitis and the presence of the HLA B27 antigen in the blood; 52% of relatives of spondylitic patients also have HLA B27 antigen.⁴

The onset of ankylosing spondylitis is insidious in 82% of the patients. Recurrent low back pain and stiffness are universal complaints, usually more severe on waking in the morning or after prolonged inactivity. In about 10% of the patients sciatic pain may be the initial symptom but usually not accompanied by neurologic abnormalities. In roughly 20% of patients the initial manifestation may be painful swelling in one or more peripheral joints or inflammation in the eyes. Fever, fatigue, loss of appetite, and weight loss may accompany an occasional severe case. Usually the sacroiliac joints are involved first, and the symptoms may be confined to this

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area for months or even years before other spinal segments are involved. Ankylosing spondylitis may vary in severity and progression. Although the inflammatory process may be acute, subacute or chronic in any or all vertebral regions, the tendency is toward a gradual spread cephalad. Thus, in patients in whom the presenting symptoms are localized to higher segments, the SI joints are usually involved at the time of examination despite absence of clinical findings there.

The physical signs of ankylosing spondylitis vary with the severity and duration of the disease and the level of spine involvement. In mild or early stages, the physical findings may be normal. More severe illness may manifest systemic changes such as muscle wasting, anemia, weight loss, and general debility. In an established case of ankylosing spondylitis the patient assumes a characteristic posture; the head is thrown forward on the shoulders, the neck carried in a rigid attitude; there is flattening of the anterior chest, kyphosis of the thoracic spines and flattening or disappearance of the normal lumbar lordosis. The pelvis is tilted forward, and there might be flexion contractures of the hips. Tenderness to palpation along the rib articulation is evident, along with decreased chest expansion. Hips and shoulders are involved in approximately 20% of cases, but involvement of the smaller joints such as elbows, wrists, hands and ankles is much less common. Inflammatory disease of the eyes (iritis or uveitis) occurs in 15% of patients at some time during the course of their disease. It may even precede the onset of sacroiliac or spinal involvement. Aortic insufficiency secondary to aortitis with widening of the aorta may be associated with abnormal heart rhythms, angina pectoris or even congestive heart failure. Pericarditis has been reported.⁵ Destructive granulomatous lesions between vertebral bodies have been described and may provide a relatively weak point in an otherwise rigid spine, predisposing the patient to fractures or cord damage, sometimes resulting in paralysis below the level of the lesion.⁶

Laboratory Tests

With the exception of x-rays and more recently HLA B27 determination, laboratory tests are not specific for ankylosing spondylitis. Of the many HLA antigens, it has been found that a particular antigen of the human leukocyte, called HLA B27, is seen with very high frequency in ankylosing spondylitis; 88-97% of patients with this disease have HLA B27 in their blood.⁷ In contrast, only 10% of the normal Caucasian population and 4% of the normal Afro-American population have the antigen. Interestingly, 52% of first degree relatives of patients with ankylosing spondylitis are + for B27, which might explain its strong familial tendency as well as the fact that it is not as common among blacks.⁷ The B27 antigen seems to be associated with the x-ray

finding of sacroiliitis more than with the other manifestations of the disease. The current feeling is that it is a specific genetic marker for ankylosing spondylitis (sacroiliitis) and may help identify those individuals who are likely to have the disease, even though they may not yet have radiographic changes of ankylosing spondylitis. Of those with positive HLA B27, the frequency of the disease has been calculated at 20-25%.

Radiologic changes are diagnostic of ankylosing spondylitis. The earliest findings are generally seen in the sacroiliac joints with blurring and irregularities of the articular margins and subchondral sclerosis adjacent to the sacroiliac joints. As the disease progresses, the joint spaces become narrowed, and bony ankylosis eventually occurs. In the spinal column the first changes are generally believed to occur in the apophyseal joints with blurring, sclerosis and narrowing of the articular margins. Anterior spondylitis or inflammation of the soft tissues anterior to the nucleus pulposus results in sclerosis and rounding of the anterior vertebral corners. There is also loss of the normal concavity of the vertebral bodies, which results in "squaring" of the vertebrae. Ossification of the paravertebral tissues follows the anterior spondylitis and syndesmophytes form between adjacent vertebral bodies. These eventually bridge over the intervertebral spaces, leading to the typical "bamboo spine appearance." Complete fusion of the posterolateral masses may occur at the cervical levels. Erosions at the margins of the symphysis pubis may occur and periosteal new bone formation may be seen along the borders of the pelvic bones, particularly at the tendinous insertions. Osteoporosis is a regular finding in established ankylosing spondylitis. Destructive granulomatous vertebral lesions may resemble metastatic cancer, infections or multiple myeloma, a malignant disease of bone marrow cells.

Differential Diagnosis

Other conditions are known to result in a stiff spine resembling ankylosing spondylitis. A severe form of degenerative osteoarthritis known as ankylosing hyperostosis of the elderly is characterized by uneven massive bony bridges between vertebral bodies, but the absence of sacroiliac joint changes distinguishes this condition from ankylosing spondylitis. Bony bridging of vertebral bodies may also be seen in hypoparathyroidism, ochronosis and familial paravertebral calcification, which are distinguished from ankylosing spondylitis by the absence of sacroiliitis and the lack of HLA B27 antigen.

An interesting group of patients with Reiter's syndrome, psoriasis, inflammatory bowel disease, and some cases of juvenile rheumatoid arthritis share certain features in common with ankylosing spondylitis, that is, they may have

sacroiliitis, spondylitis and positive HLA B27.⁸ In these instances, the clinical manifestations of the primary disease help to classify it in its proper perspective.

In summary, ankylosing spondylitis is a chronic inflammatory disease of man characterized clinically by progressive stiffening of the back and pathologically by bony fusion of all or parts of the axial skeleton including the sacroiliac joints. Diagnosis may be confirmed by characteristic radiologic changes and the presence of HLA B27 antigen in the blood.

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