A Retrospective Study of a Skeleton Featuring Ankylosing Spondylitis (AS)

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Abstract : **The skeleton of a Thai male teacher who died at the age of 75 was studied. The skeleton showed severe spinal fusion and deformities. We studied the pathological features and hypothesized a diagnosis through a retrospective study of the case history. Pathological findings showed severe spinal ankylosis due to continuous fusion of the vertebral bodies from T2 to L3 and L4 to L5, the apophyseal joints from T2 to L1, the sacrococcygeal and atlantooccipital joints, and ankylosis of the bilateral sacroiliac joints. Ossification of the anterior longitudinal ligaments from T2 to L3 and the supraspinous ligament from T6 to T12 and L4 to L5 were observed. Kyphosis and costovertebral ankylosis which obliterated the intervertebral foramina were found at T7, T8 and T9. Synostosis of C2 to C3 and ossification of the right sacrotuberous ligament were observed. Enthesopathy was seen in the sternocostal radiate ligaments and also the lower limb ligaments and tendons. These features are characteristic of severe progressive ankylosing spondylitis. The case history recorded no report of clinical manifestations of spinal problems. The patient was healthy until the last 2 to 3 years of his life, when he exhibited difficulty in leaning sideways, breathing, and had chest pain. He was diagnosed and treated for ischemic heart disease but suffered from breathing difficulties until he died. He was never diagnosed with ankylosing spondylitis.**

Key words: ankylosing spondylitis, spine ankylosis, calcific enthesopathy

เรื่องย่อ : **การศึกษาย้อนอดีตของโครงกระดูกที่มีลักษณะผิดปกติเฉพาะแบบ ankylosing spondylitis (AS)**

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ผู้วิจัยค้นพบโครงกระดูก 1 โครงที่มีลักษณะผิดปกติอย่างรุนแรงของชายไทยอายุ 75 ปี อาชีพครู ซึ่งก่อให้เกิดความสนใจ และตั้งวัตถุประสงค์ที่จะศึกษาลักษณะของพยาธิสภาพ และตั้งสมมติฐานของโรคที่ก่อให้เกิดความผิดปกติในโครงนี้ ตลอดจนศึกษาประวัติอดีตและความเจ็บป่วยขณะยังมีชีวิตอยู่ว่าเป็นอย่างไร โดยเฉพาะความผิดปกติในการเคลื่อนไหว โครงกระดูกแสดงลักษณะผิดปกติคือมีการติดแข็งของลำกระดูกสันหลังจากการเชื่อมติดของ vertebral bodies ตั้งแต่ระดับ T2-L3 และ L4-L5 มีการติดเชื่อมของข้อต่อต่าง ๆ ของกระดูกสันหลัง ได้แก่ ข้อต่อ apophyseal ตั้งแต่ระดับ T2-L1, ข้อต่อ sacrococcygeal, ข้อต่อ atlantooccipital, และข้อต่อ sacroiliac ทั้งสองข้าง มีการพอกของแคลเซียมบนเอ็น anterior longitudinal เป็นลำยาว ตั้งแต่ T2-L3 และบนเอ็น supraspinous ตั้งแต่ T6-T12 และ L4-L5 มีการติดเชื่อมของหัวกระดูกซี่โครงกับกระดูกสันหลัง T7, T8, และ T9 ทำให้ปิดรูทางออกของรากประสาทระหว่าง T7-T8 และ T8-T9 มีการเชื่อมต่อสนิทแบบ synostosis ระหว่างกระดูกสันหลัง C2-C3 ลำกระดูกสันหลังโค้งงอแบบหลังโกง เอ็น sacrotuberous ข้างขวากลายเป็นกระดูก มี calcific enthesopathy ที่ข้อต่อ sternocostal และปลายเอ็นต่าง ๆ ที่เกาะกระดูกรยางค์ล่าง ลักษณะผิดปกติทั้งหมดดังกล่าวเป็นลักษณะของโรค ankylosing spondylitis เมื่อติดตามประวัติย้อนหลังตอนยังมีชีวิตอยู่ พบว่าไม่เคยไปพบแพทย์ด้วยปัญหาเกี่ยวกับกระดูกหรือเส้นประสาทสันหลัง จนกระทั่งเมื่อ 2-3 ปีก่อนเสียชีวิต จึงเคลื่อนไหวลำบากและนั่งพับเพียบไม่ได้ ไปพบแพทย์บ่อยมากด้วยเรื่องหายใจลำบากและเจ็บแน่นหน้าอก ได้รับการวินิจฉัยว่าเป็นโรคหัวใจและได้รับการรักษาแบบโรคหัวใจโดยไม่เคยได้รับการวินิจฉัยว่าเป็นโรค ankylosing spondylitis เลย

INTRODUCTION

Ankylosing spondylitis (AS; also called Marie-Strumpell disease or von Bechterew's disease) is a seronegative (rheumatoid factor negative) arthritis affecting the sacroiliac, vertebral and costovertebral joints and may involve peripheral joints.[1,2] The prevalence is 0.5-2%.[1,3] The etiology is unknown but the disease is frequently associated with HLA-B27[1] and males are more commonly affected than females.[3] Radiographic evidence of sacroiliitis is the most specific criteria for AS diagnosis. Pathological findings identified by radiographs show squaring of the vertebral bodies, bridging with syndesmophytes, calcification of the annulus fibrosus (bamboo spine) and ankylosis of the sacroiliac joints. Reports of a skeleton with AS are rare so when we found a skeleton with severe spinal fusion and deformities featuring AS, we investigated it.

MATERIALS AND METHODS

Among 110 dried skeletons at the Department of Anatomy, Faculty of Medicine, Khon Kaen University, we observed one with severe spinal fusion and deformities. The pathological features were studied and described. The case history was studied retrospectively by interviewing the patient's son and co-workers. Clinical records from the hospital which followed him up for 2½ years before his death were studied with the permission of the son and the hospital director.

RESULTS

The skeleton was from a Thai male teacher who died at the age of 75. He was a football player and healthy until middle age, when he started to complain of backaches though he never considered these serious. The clinical reports recorded no back pain, motor disabilities or any other underlying disease. He retired at 62. Thirteen years later, two years before he died, he had difficulty in leaning sideways, breathing, and suffered chest pains. He was diagnosed and treated for ischemic heart disease with breathing insufficiency because a worsening problem.

Pathological findings included continuous fusion of the vertebral bodies T2 to L3 and L4 to L5 (Figure 1), the apophyseal joints from T2 to L1

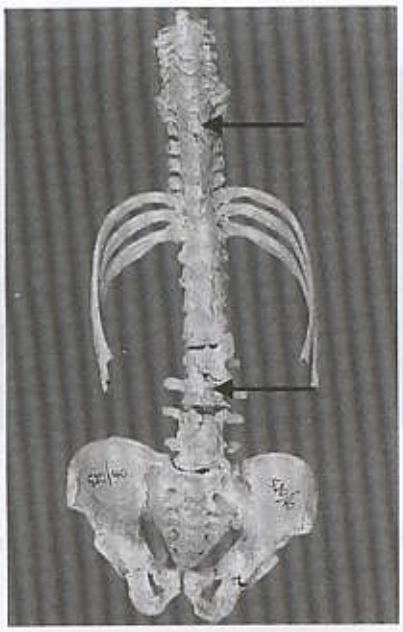

Figure 1. Fusion of vertebral bodies from T2 (upper arrow) to L3 (lower arrow), and L4 to L5.

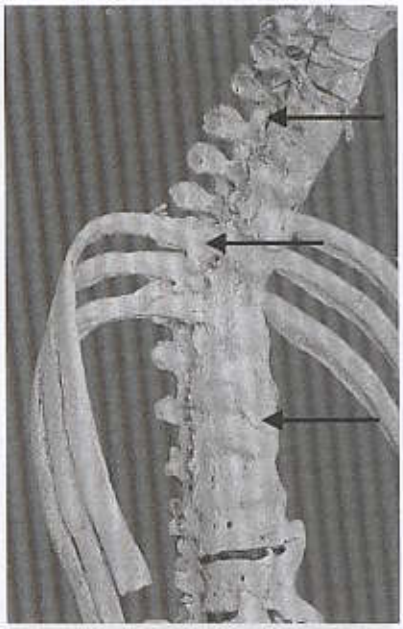

Figure 2. Fusion of the apophyseal joint (upper arrow), costovertebral ankylosis (middle arrow), and ossification of the anterior longitudinal ligament (lower arrow).

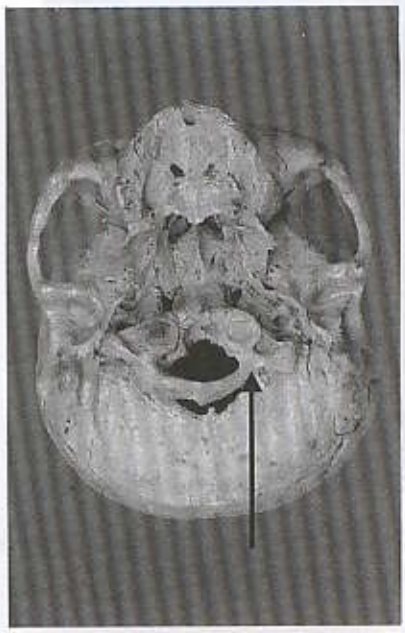

Figure 3. Fusion of the atlantooccipital joint.

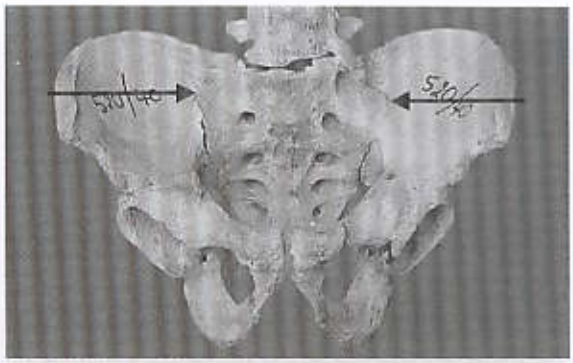

Figure 4. Bilateral ankylosis of the sacroiliac joints.

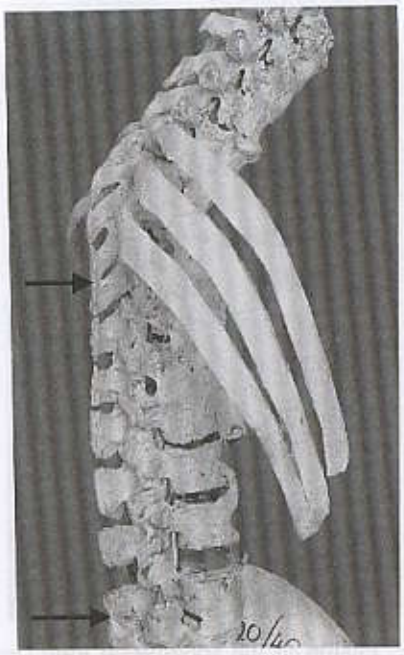

Figure 5. Ossification of the supraspinous ligament in thoracic region (upper arrow) and the interspinous ligament in lumbar region (lower arrow).

Figure 7. Synostosis of the second and third cervical vertebrae.

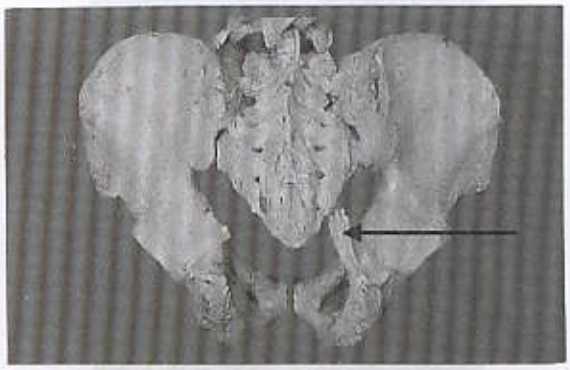

Figure 6. Ossification of the right sacrotuberous ligament.

Figure 8. Calcific enthesopathy and ankylosis of the sternocostal joints.

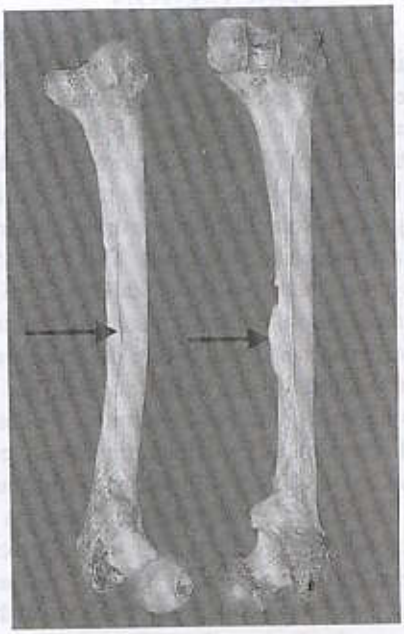

Figure 9. Calcific enthesopathy at the linea aspera of the femur.

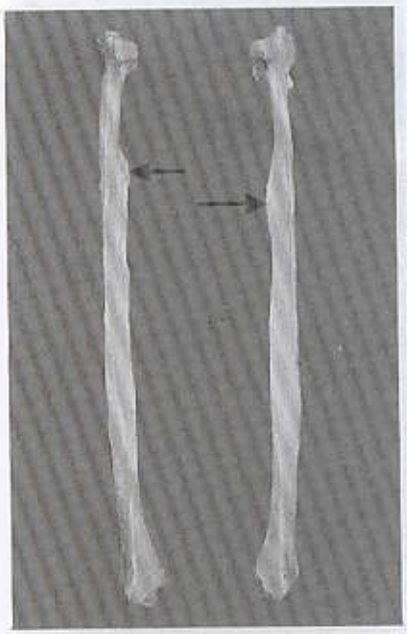

Figure 10. Calcific enthesopathy at the interosseous border of the fibula.

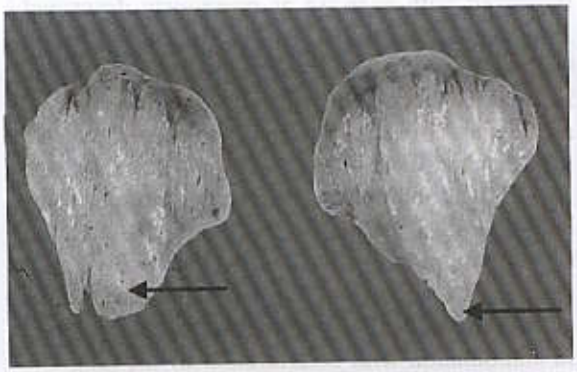

Figure 11. Calcific enthesopathy of the patellar ligament.

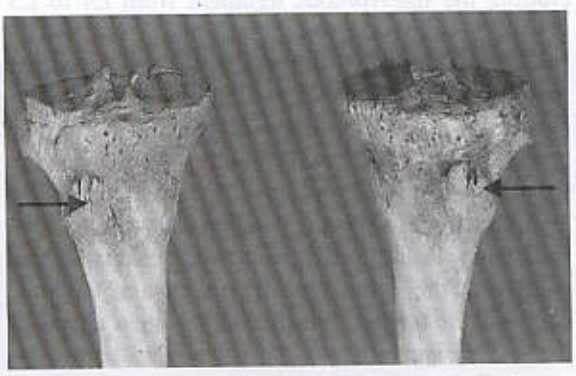

Figure 12. Proximal tibia showing calcific enthesopathy of the patellar ligament.

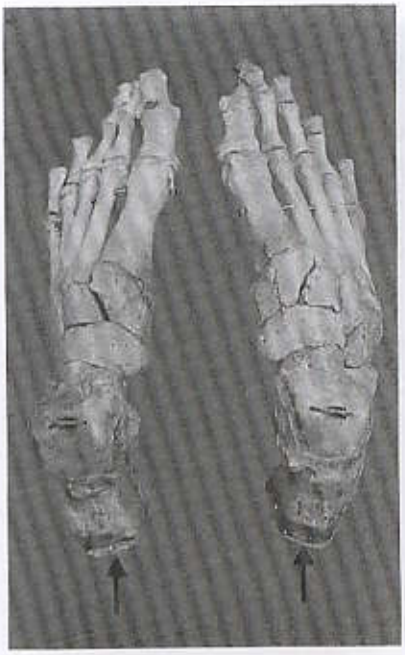

Figure 13. Calcific enthesopathy of the tendocalcaneus resulting in a calcaneal spur.

(Figure 2), the atlantooccipital joint (Figure 3) and the sacrococcygeal joints, and bilateral ankylosis of sacroiliac joints (Figure 4). Ossification of the anterior longitudinal ligament from T2 to L3 (Figure 2) and the supraspinous ligament from T6 to T12, including the interspinous ligament from L4 to L5 (Figure 5) were observed. Kyphosis and costovertebral ankylosis which obliterated the intervertebral foramina were found at T7 to T9 (Figure 2). Ossification of the right sacrotuberous ligament (Figure 6) and synostosis of C2 to C3 (Figure 7) were observed. Ankylosis of the sternocostal joints due to calcific enthesopathy of the radiate ligaments could be seen (Figure 8). Additionally, calcific enthesopathy was evident in the lower limb ligaments and tendons, namely: the tendons attached to the linea aspera of the femur (Figure 9), the tibiofibular interosseous ligaments (Figure 10), the patellar ligaments (Figures 11 and 12) and the calcaneal ligaments. The latter calcific enthesopathy caused a spur of the heel (Figure 13). The foregoing features are all characteristic of severe progressive AS.

DISCUSSION

The typical pathology of AS involves the sacroiliac, vertebral and costovertebral joints especially the thoracolumbar spine, and may involve the peripheral joints.[1,2] Calcific enthesopathy at the junction of the annulus fibrosus and vertebral end plates leads to bridging by syndesmophytes.[4-7] As the disease progresses, some patients develop a decreasing lordosis and increasing kyphosis. This skeleton presents the complete pathological features of AS which we have rarely seen. Moreover, some of the findings have never been described before, such as fusion of the atlantooccipital joint, ossification of the sacrotuberous ligament, and the obliteration of the intervertebral foramina. The deformities are severe and must have led to difficulty in motor performance.

The Rome criteria for the diagnosis of AS include clinical and radiologic criteria. The clinical criteria consist of low back pain and stiffness for more than three months, pain and stiffness in the thoracic region, limited motion of the lumbar spine, limited chest expansion and a history or evidence of iritis or its sequelae. Radiologic criteria include bilateral sacroiliitis.[1] AS is diagnosed if sacroiliitis presents with any one of these clinical criteria.

Surprisingly, the patient had not presented to the doctor any clinical symptoms of AS nor demonstrated any motor problems until just a few years before death. Possibly, as a physically active person and sport player, his symptoms were relieved and improved with exercise as previously described.[1] However, at the end of his life he would not have been as active; indeed motor activity was restricted. He could not lean sideways. This could be due to the ankylosis of bilateral SI joints, fusion of the lumbar spine, and ossification of the sacrotuberous ligament. He still was able to walk since both hip joints were not involved. The thoracic cage was rigid due to costovertebral and sternocostal ankylosis and vertebral fusion. The rigidity of the thoracic cage reduced chest movement. The reduction in chest movement in combination with deterioration of the lung elasticity due to increasing age seriously limited the respiratory volume. This resulted in progressive breathing insufficiency. He was diagnosed and treated for ischemic heart disease, but never for AS.

Hochberg, et al reported a group of AS patients with painless spinal ankylosis and reduced chest expansion.[8] These patients were classified as a "latent" form of AS with silent axial disease. Therefore, AS patients may present with breathing insufficiency, which may be misdiagnosed or overlooked if radiographs are not investigated.

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