

## A Case of Descending Thoracic and Abdominal Aortic Aneurysm with Ankylosing Spondylitis

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Ankylosing spondylitis (AS) is a systemic inflammatory disorder that affects the axial skeleton. It often involves the extra-articular organs. Cardiovascular involvement is one of the extra-articular manifestations, which is mostly represented by aortic root, valvular heart disease, and conduction disturbances. An aortic sclerosing inflammatory process induces aortic root thickening and rigidity. An aortic aneurysmal change is a rare complication that often

leads to life threatening conditions. A few cases regarding aortic aneurysm have been reported, but there are no reported cases in Korea. We report the first case of descending thoracic and abdominal aortic aneurysm in a patient with ankylosing spondylitis.

**Key Words.** Ankylosing spondylitis, Extra-articular manifestation, Cardiovascular, Aortic aneurysm

### Introduction

Ankylosing spondylitis (AS) is the most common and severe subtype of spondyloarthritides that primarily affects the axial skeleton, most the sacroiliac joints. It often involves the extra-articular organs represented by anterior uveitis, inflammatory bowel disease, lung and kidney abnormalities, osteoporosis, and psoriasis etc. An epidemiological study of 847 patients in Belgium found that 42% had one or more extra-articular manifestations (1).

Cardiovascular involvement is well known extra-articular manifestation of AS. The usual manifestations such as aortic valve insufficiency, conduction abnormalities, and left ventricular hypertrophy (LVH) may occur alone or together.

A few cases about complications such as aortic aneurysm and bicuspid valve have been reported abroad, but there is no report in Korea. Here we report a case of a patient with descending thoracic and abdominal aortic aneurysm without any other cardiovascular involvements.

### Case Report

A 59-year-old male patient with 13-year history of AS presented with abdominal aortic aneurysm detected incidentally during the annual medical check-up. He was initially diagnosed with AS on December, 2001, complaining of inflammatory back pain. He had limitation of chest expansion and spinal flexion. Pelvic plain x-ray and magnetic resonance imaging showed active sacroiliitis (Figure 1). Erythrocyte sedimentation rate and C-reactive protein was 58 mm/h and 11 mg/dL. HLA-27 test was negative. He was complicated with anterior uveitis and osteoporosis. Other extra-articular manifestations were not combined. He was treated with non-steroidal anti-inflammatory drugs (NSAIDs), sulfasalazine, methotrexate and tumor necrosis factor inhibitor, etanercept (enbrel<sup>®</sup>), was started due to incomplete treatment response since 2004. Recent Bath Ankylosing Spondylitis Disease Activity Index (BASDAI) was 2.55 points.

During the annual medical check-up, the abdominal aortic aneurysm was detected by abdominal computed tomography

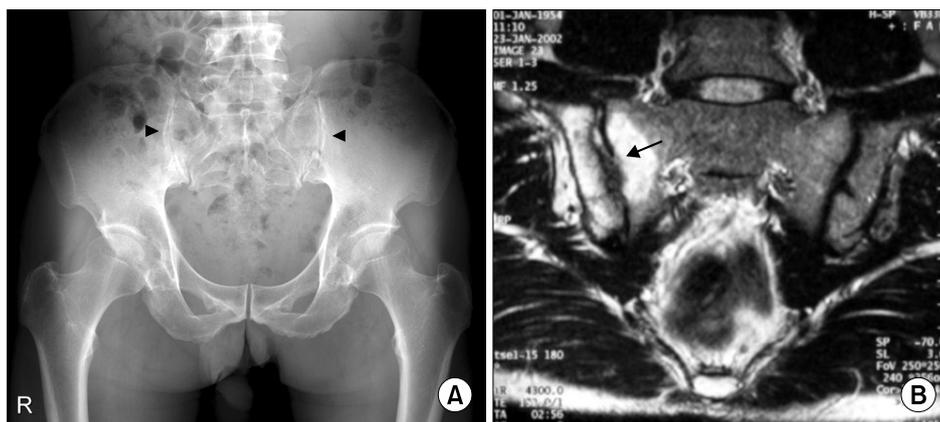
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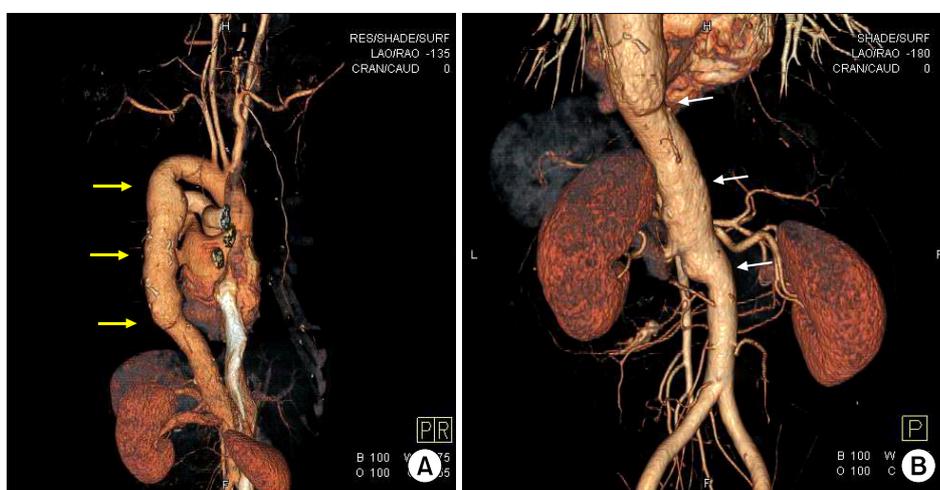
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**Figure 1.** Sacroiliitis at initial presentation. (A) Subchondral sclerosis with periarticular erosions were observed at both sacroiliac joint in plain pelvic X-ray. Narrowing of the sacroiliac spaces also could be seen (arrowheads: Sacroiliitis, New York classification III). (B) Pelvic magnetic resonance imaging showed bone marrow edema and joint space narrowing in right sacroiliac joint. Also synovial enhancement and subchondral erosions were seen (arrow).



**Figure 2.** Incidentally detected aortic aneurysm. (A) Chest computed tomographic imaging of three-dimensional reconstruction shows a dilated descending aorta (yellow arrows). Aortic root and aortic arch appeared to be normal. (B) Continuing abdominal aortic aneurysm involves the suprarenal and infrarenal segment of abdominal aorta (white arrows).

unexpectedly and following work-up chest computed tomography revealed additional descending aortic aneurysm. The fusiform aneurysm involved the entire descending thoracic aorta with small eccentric mural thrombus in distal portion, measuring about 4.3 cm in the diameter and total length was about 16 cm. The aortic root and ascending aorta were not involved (Figure 2A). Continuing abdominal aortic aneurysm involved the suprarenal and infrarenal segment of abdominal aorta, measuring about 3.9 cm in the diameter. The distal neck from the aortoiliac bifurcation measured about 6.7 cm in length (Figure 2B).

He did not complain any symptoms except mild chest discomfort. Electrocardiogram showed regular sinus rhythm without any conduction abnormalities. Echocardiography was followed to evaluate the possibility of other cardiovascular involvement. Both aortic valve function and ejection fraction were within normal limit. He is well tolerable without any complications at out-patient clinic.

## Discussion

The prevalence of cardiac involvement in patients with AS has been reported to be 10% to 30%. Aortic root, valvular heart disease and conduction disturbances are the most common manifestations. Generally these manifestations follow the arthritic symptoms by 10~20 years, and rarely precede it (2,3). Roldan et al reported that 82% of the AS patients had subclinical aortic root and valve disease by echocardiography (4). Aortic incompetence was noted in 3.5% of AS patients who had the disease for 15 years and in 10% after 30 years (5). Previous report that analyzed clinical profile of total 721 AS patients in Korea revealed that cardiac involvement was not common in Korea. Only 13 (1.6%) patients had cardiac involvement and 8 of those patients had cardiac conduction abnormalities (6).

In patients with AS, aortic aneurysmal dilation of thoracic or abdominal aorta is an uncommon condition that rarely has been reported. Proceeding sclerosing inflammatory process is supposed to induce aortic root thickening and aortic rigidity (7). The following histopathological features of the in-

flammation in the aorta of 8 AS patients were reported by Bulkley and Roberts in 1973: (i) Focal destruction of muscular and elastic structures of the media; (ii) thickening of intima and adventitia; and (iii) obliterative vessel disease (8). This aortitis leads to aortic root dilatation and aortic valve incompetence. In addition to the aortic root, the chronic inflammation may extend into the ventricular septum and can cause conduction disturbances (9).

Yuan analyzed 17 cases of aortic aneurysm associated with AS in the English literature (10). There were 13 males and 4 females. Mean age and disease duration of the patients were 44.9 and 13.4 years, which are similar to those of our patient. However few significant differences from the previous cases are noted. Our case patient was HLA B27 negative, which is not usual pattern of AS. Out of previous 17 patients, 10 patients were not tested for HLA B27, and only one patient was tested for negative. Secondly, the ECG and echocardiography of our patient showed that heart structure and function were normal, unlike some of the previous patients who showed other cardiac manifestations; aortic insufficiency in 7 (41.2%), heart block in 4 (23.5%), and LVH in 7 (41.2%) patients. Last but not least, aneurysmal dilations prior cases were mostly in the aortic root and ascending thoracic aorta; in the aortic root in 3 (17.6%), ascending aorta in 6 (35.3%), ascending aorta and arch in 1, aortic root, ascending aorta and arch in 1 (5.9%) and entire thoracic aorta in 2 (11.8%), respectively. Our patient had descending thoracic and abdominal aorta involvement, and descending thoracic involvement occurred only in 3 patients (17.6%) of the previous cases, and only one case (5.9%) for abdominal aorta involvement.

Mortality attributable to AS is largely the result of extra-articular involvements such as spinal trauma, aortic insufficiency, respiratory failure, amyloid nephropathy, or upper gastrointestinal hemorrhage. Circulatory disease is the most frequent cause of death of patients with AS (11). Aortic involvement is a rare but often leads to life threatening condition that sometimes needs surgical approach.

In this case, we report a patient with AS who had aortic aneurysmal change of unusual site. To our knowledge, it is the first report of aortic aneurysm involving both descending

thoracic and abdominal aorta in AS patients. We should consider the possibility of aortic involvement that can affect the clinical outcomes and prognosis of AS.

### Summary

Aortic aneurysmal change is a rare cardiovascular involvement but often leads to life threatening condition in AS patient. Rheumatologists should carefully consider the possibility of this complication to provide appropriate management.

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