

CASE STUDIES

Cardiovascular manifestations of spondyloarthritis: Aortoarteritis of ankylosing spondylitis or Takayasu's aortoarteritis?

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Abstract

Ankylosing spondylitis (AS) is an autoimmune disease with various articular and extra-articular manifestations. The cardiovascular manifestations of the disease include aortitis, aortic valve disease, conduction disturbances, aortic or mitral regurgitation and cardiomyopathy. The present study deals with an unusual presentation of short clinical history of spondyloarthritis, and initial presentation of cardiovascular manifestations aortic regurgitation and aortoarteritis.

Keywords: spondyloarthritis, aortic regurgitation, cardiovascular manifestations, aortitis

Introduction

An association between spondyloarthritis (SpA) and Takayasu's arteritis (TA) has been reported in a few case series.¹ Aortic regurgitation (AR), AV blocks and other less common cardiovascular manifestations have a known association with SpA. Moreover, there is a known involvement of aortic valve, aorta and aortic arch in SpA. These findings are usually noted in SpA patients with longer disease duration and a known history of diagnosis and treatment for SpAs. The present case study deals with an unusual case of very short clinical history of low back pain and cardiovascular manifestations (AR + aortitis) as initial presentation.

Case report

A 27-years-old female patient presented for a cardiology consultation with chief complaints of progressive shortness of breath for last 6 months and chest pain for last 3-4 months. Over the past 1 month, the gradual onset exertional dyspnea progressed from NYHA grade 2 to grade 3. She had episodes of paroxysmal nocturnal dyspnea. The chest pain, localized in the epigastric region, increased after exertion and did not show diurnal variation. The pain was not radiating to left arm or other sites. Other complaints included cough with expectoration and transient swelling of feet, which relieved with rest. There was no history of limb claudication, headache, and visual disturbances. The 2D-echo findings revealed aortic regurgitation (AR).

Repeat echo was suggestive of severe AR, moderate tricuspid regurgitation (TR), moderate left ventricular (LV) dysfunction and low ejection fraction (EF) of 40%.

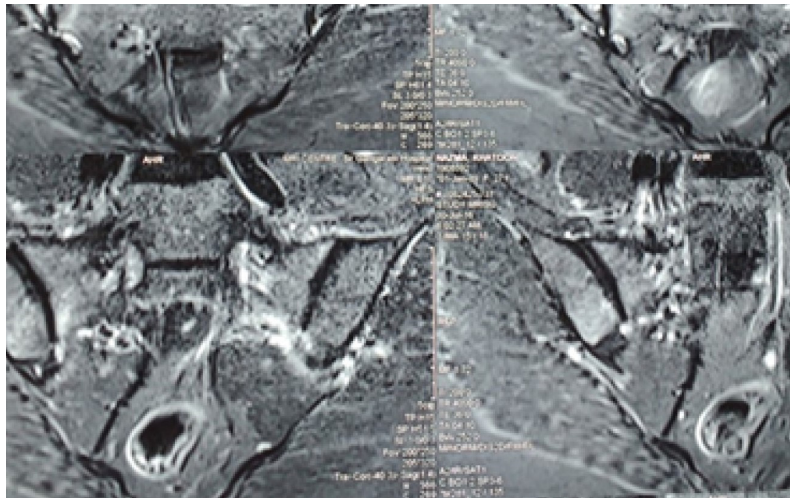
The patient was referred to consult a rheumatologist to rule out other causes of AR, as 2D-echo was not suggestive of rheumatic heart disease. Elaborative history revealed presence of inflammatory low backache for the past 5-6 months. There was no history of eye, skin or gut problems. Blood pressure readings noted in right and left arm were 112/60 mmHg and 100/56 mmHg respectively. Lower limb blood pressure noted in right and left lower limb were 140/84 mmHg and 138/86 mmHg respectively. Pulse was 88/min with high in volume and symmetrical in both upper limbs. There was no radiofemoral delay, and pulses in lower limbs were equally palpable with normal force. There abdominal or carotid bruits were absent.

General examination was normal with normal jugular venous pressure. Marfanoid features were absent. Systolic murmur was present in mitral area, aortic area, and 3rd left parasternal area. She had peripheral signs of AR. Musculoskeletal examination indicated FABER test was positive bilaterally. Modified Schober's test showed 4.5 cm expansion suggestive of lumbosacral restriction. Lateral flexions at lumbar spine on right and left side were 16 and 18 cm respectively. Cervical rotation was 100° bilaterally. Tragus-to-wall distance and intermalleolar distance were

Fig. 1: X-ray of pelvis (AP) suggestive of bilateral sacroiliitis (right more than left)



Fig. 2: MRI sacroiliac joints suggestive of right sided more than left sided active sacroiliitis



7.9 cm and 128 cm respectively.

Her ESR was 79 mm/ fall in 1st hour and CRP was 68 mg/L. X-ray of pelvis showed right sided grade 3 and left sided grade 2 sacroiliitis (Fig. 1). MRI of both the sacroiliac joints (STIR images) showed active sacroiliitis (right>left) (Fig. 2). HLA-B27 test was positive. She was also investigated for mycobacterium due to decreased appetite. CT chest/ abdomen showed diffuse thickening of almost entire aorta with the thickening involving the origin of the branches of the aortic arches consistent with an arteritis, possibly Takayasu's arteritis (Fig. 3a and 3b). Mantoux test was negative. Coronary angiography findings were normal.

In view of symptomatic severe AR with decreased ejection

fraction, the patient had undergone aortic valve repair. Biopsy of valve and wall of ascending aorta (Fig. 4a, 4b, & 5) was suggestive of non-specific aortitis with moderate thickening of adventitia and intima and myxomatous degeneration of the aortic valve.

In view of cardiac surgery, NSAIDS and steroids were deferred juxta-operatively. Mild bearable backache was managed conservatively with physiotherapy and exercise program. Sulfasalazine and NSAIDS (Tablet naproxen) were prescribed after 1 month of surgery. ESR and CRP noted during follow-up were 68 mm and 40 mg/L respectively. Thoracic and abdominal CT angiography was done after 1 month of sulfasalazine and naproxen. Thickening of aorta and its branches, without any stenosis or aneurysmal

Fig.3a & 3b: Thickening of aorta and its branches at their origin

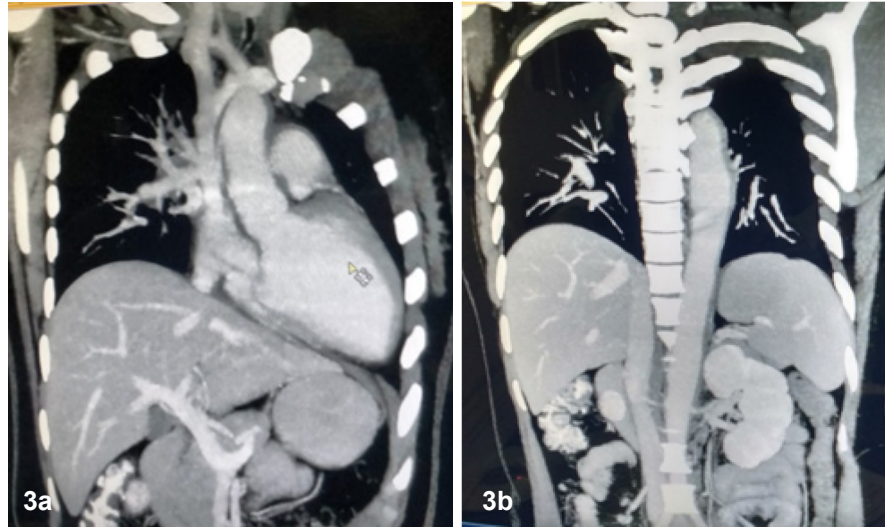


Fig. 4a & 4b: Biopsy of aortic valve showing lymphocytic infiltration, more in intima and myxomatous degeneration of valve substance

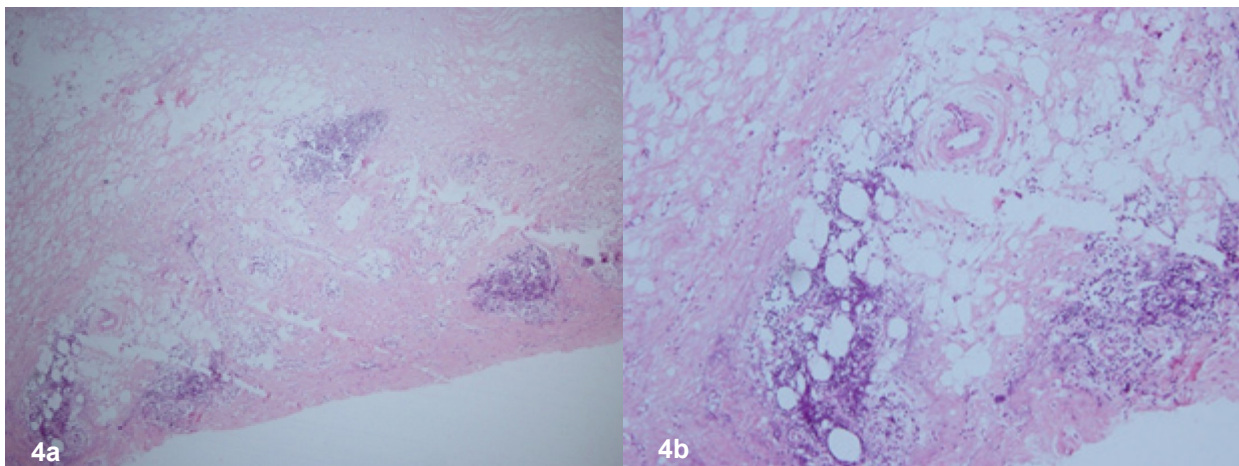
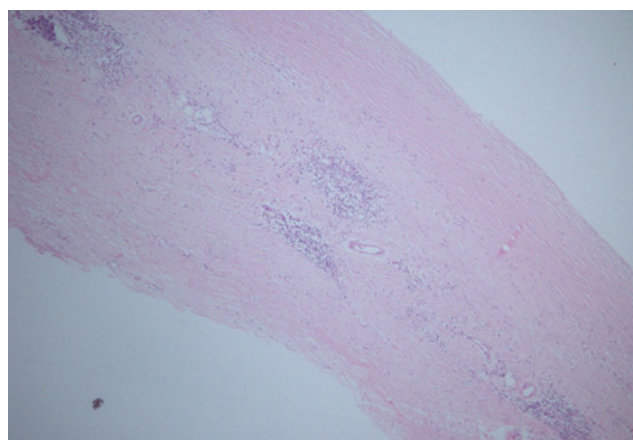


Fig. 5: Biopsy of aortic wall showing moderate thickening of adventitia and intima, and mononuclear cell infiltrates in all three layers of vessel wall (more in media)



dilatation, was diminished in size when compared to previous scan (Images not available). Prednisolone 30 mg/day was started after two months of surgery, as she was asymptomatic and there was radiological improvement in aortoarteritis. Prednisolone had been tapered in next 3 months and she is currently on prednisolone 5 mg/day as third CT angiography revealed similar changes, no progression and last CRP level was 16 mg/L.

Discussion

Occurrence of cardiac manifestations in ankylosing spondylitis is well described, with cardiac conduction abnormalities (AV blocks and intraventricular blocks) being the most common. Other typical abnormalities are aortic regurgitation (AR), aortitis, ischemic heart disease, peripheral vascular disease and heart failure. Likelihood of these conditions increases with duration of the ankylosing spondylitis. HLA-B27 is a stronger risk factor for cardiac manifestations of ankylosing spondylitis.

AR in ankylosing spondylitis can be easily differentiated from AR associated with other cardiac disorders. This is characterized by typical dilatation of aortic root, fibrosis, thickening and downward pull of the bases of the cusps with inward rolling of the edges/margins of the cusps. The term 'subaortic bump' refers to fibrosis of the base of the anterior mitral leaflet.²

Focal destruction in media and thickening of adventitia and intima is noted in ankylosing spondylitis patients with AR. There are no reports on the prevalence of aortitis in ankylosing spondylitis, whereas the prevalences of aortic valve disease in early AS and in later disease stages have been estimated to be 4% (a disease of < 15-year duration) and 10% (a disease of >30 years), respectively.³

A study by Labresh *et al.*, involving 36 SpA patients, has found subaortic fibrosis and leaflet thickening in 30.5% patients. All the patients had longer disease duration (28.1 versus 17.7 years). They found that significant percentage of patients with SpA have aortic root disease before clinical onset of AR.⁴

Another study conducted on 44 ankylosing spondylitis patients has demonstrated that aortic root and valve disease were common in 82% of the subjects. Among these subjects, 50% had AR and 40% had moderate lesions. The study has found that only duration of disease is related to AR and it is unrelated to disease activity, severity and therapy.

Around 20% of the subjects developed heart failure and had undergone valve replacement.⁵

Bergfeldt *et al.* have evaluated 91 patients with lone AR to study the possible role of HLA-B27 in the development of lone AR. The study has found that HLA-B27 associated inflammatory disease might be the cause for AR in around 15-20% of patients, but no significant association was found between lone AR and HLA-B27. On the other hand, HLA-B27 was noted in 88% of the male patients with combination of AR and conduction abnormalities.⁶

Apart from Takayasu's arteritis (TA), other autoimmune diseases that can affect aorta include giant cell arteritis, Cogan's syndrome, relapsing polychondritis, rheumatoid arthritis, Behcet's disease and SpA. The present study raises a significant question: what terminology should be used for signifying aortoarteritis associated with other autoimmune diseases, Takayasu's arteritis or secondary aortoarteritis? Disease monitoring and treatment would have major impact on patient's life, as concluding the diagnosis influences the prognostic course. Hence the clinicians should exercise utmost caution while deciding the strategy for managing underlying autoimmune disease associated with aortoarteritis.

Conclusion

There is no prevalence data on AR and aortitis in patients with SpA. HLA-B27 is associated with AR and SpAs, and only duration of disease affects prevalence of AR and other cardiovascular manifestations of SpAs. The current case, dealing with heart failure and very short history of clinical disease of SpA (06 months), is very rare. The patient also had asymptomatic aortitis, as she did not have hypertension or other features of aortitis. HLA-B27 association has been noted in male patients with SpAs and AR plus conduction defects. The present study is first of its kind that have shown similar association in a female patient.

Competing interests

The authors declare that they have no competing interests.

Citation

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