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Case Report
Large Vessel Vasculitis

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Large Vessel Vasculitis in a Case Of Hlab27 Associated Axial Spondyloarthritis

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A 24-year-old male, with a history of inflammatory back pain of 6 years, diagnosed with Radiographic axial spondyloarthritis, using alternate therapies, presented with incidentally found discrepancy of pulses and blood pressure in upper and lower extremities on follow-up. Spondyloarthritis disease burden was high with elevated inflammatory markers, and high disease activity was observed.

Imaging of blood vessels with CT aortography revealed thrombotic occlusion of upper limb arteries and stenosis of large vessels. Evaluation for the underlying procoagulant state was negative. FDG-PET CT scan revealed metabolically active vasculitis involving ascending, arch, and descending aorta, and brachiocephalic trunks with occlusive thrombosis of carotid and upper limb vessels. Concomitant occurrence of Takayasu arteritis with Ankylosing Spondylitis was contemplated, considering several reported associations of the two disorders in literature. However, owing to the chronic underlying Axial Spondylitis with high disease activity, spondyloarthritis-associated aortitis was considered. High-dose glucocorticoids were administered, and Anti TNF agents were initiated after latent TB prophylaxis.

The presence of high inflammatory markers in Spondyloarthritis should alert one to look for extra musculoskeletal manifestations in Spondyloarthritides in the form of vasculitis.

Keywords: Spondyloarthritis, large vessels vasculitis, vasculitis

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Introduction

Radiographic axial spondyloarthritis (earlier Ankylosing spondylitis) is a chronic systemic inflammatory disorder with involvement of spine and sacroiliac joints in young males. Aortic involvement is an uncommon extra-articular manifestation of spondyloarthritis (SpA), mainly axial spondyloarthritis. It usually presents with aortic valve regurgitation (AR) secondary to inflammation and dilation of the aorta and can rarely cause aortitis, peri aortitis, or conduction abnormalities. Aortic involvement in axial spondyloarthritis is usually described as a complication of longstanding disease. It has rarely been described in the first decade of disease. We report the case of a young male with axial spondyloarthritis with poorly controlled disease complicated by mobility restriction incidentally to have discrepant pulse and blood pressure in extremities was worked accordingly and found aortitis presenting with pulse and blood pressure discrepancy in upper extremities.

Case Report

A 24-year-old male, with 6 years of inflammatory back pain peripheral arthritis of knee joints, and Achilles tendinitis had restricted spine mobility on examination. On follow-up examination, he was incidentally found to have absent pulses in the upper extremities and a discrepancy in blood pressure between the upper and lower extremities. Mean Arterial pressure in upper limbs was 70mmHg, and MAP in lower limbs was 156mmHg. His history is significant for non-compliance to allopathic treatment and persistently high inflammatory markers. The evaluation showed restriction of movements in the cervical and lumbar spine. Occiput wall distance 7cm, tragus wall distance 12cm, chest expansion 2cm, modified Schober's test 2cm, lateral lumbar flexion 2cm, intermalleolar distance 85cm, restricted cervical flexion and extension. Entheseal sites normal. Radiography revealed bilateral grade 4 sacroiliitis (Figure 1,2). Inflammatory markers ESR 45mm/hour (0-15mm/hr in men), CRP 26mg/dL (0-5mg/dL). HLA B27 by RT-PCR was positive. Disease burden was high with BASDAI (Bath Ankylosing Spondylitis Disease Activity Index) 4.6 (range 0-10), BASMI (Bath Ankylosing Spondylitis Metrology Index) 5.8 (range 0-10)

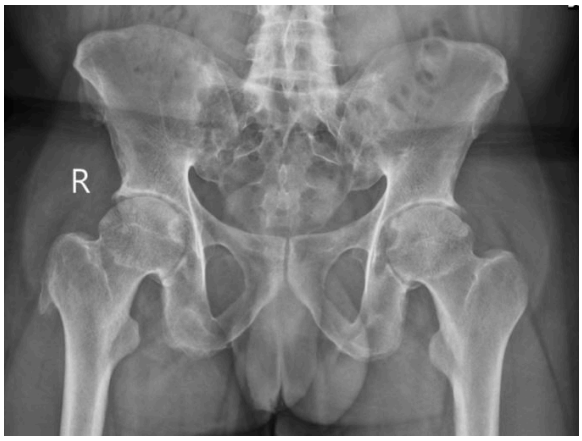

Figure 1: Bilateral grade 4 sacroiliitis

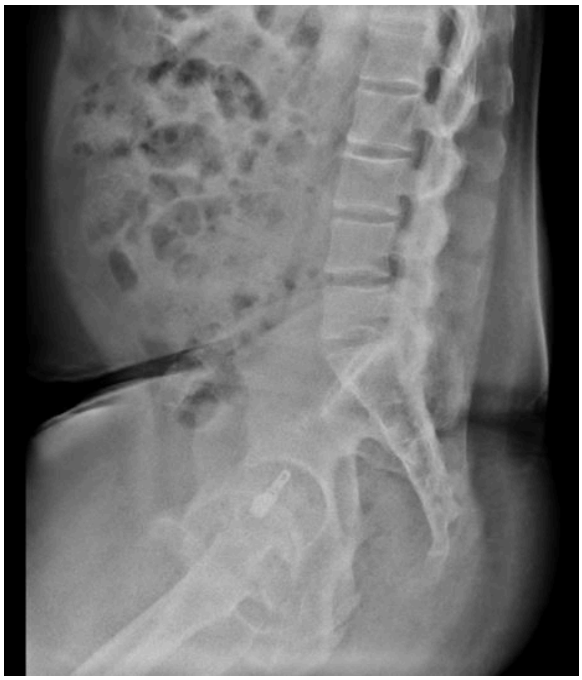

Figure 2: Squaring of vertebrae, straightening of spine

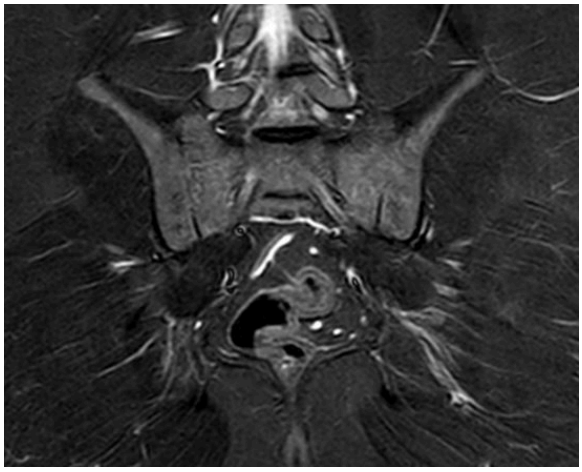

Figure 3: MRI showing ankylosed Sacroiliac joints

Due to the absence of pulses in the upper limbs with discrepancy in blood pressure in the extremities, with upper extremities showed blood pressure of 74/52mmHg, lower extremities showed 217/130 mmHg. Suspicion of large vessel vasculitis with or without thrombosis was considered.

On evaluation for the cause of absence of pulses, CT angiography of upper limb vessels was done which showed thrombotic occlusion in bilateral common carotid arteries, and bilateral subclavian and left axillary arteries. Stenosis of the brachiocephalic trunk was noted. Tests for underlying prothrombotic states were negative – Antiphospholipid antibodies (Anticardiolipin, Beta 2 glycoprotein, Lupus anticoagulant) negative, serum lipid levels normal, serum homocysteine level normal. Anti-nuclear antibodies by immunofluorescence negative.

PET CT scan was done because of large vessel stenosis, to look for inflammation of vessels. It showed metabolically active foci of uptake in ascending aorta, arch, bilateral brachiocephalic trunk, descending thoracic and abdominal aorta up to bifurcation suggestive of active vasculitis. Metabolic activity was noted at the anterior end of the transverse process of the C6 vertebra and anterior end of the right 2nd rib with slight surrounding inflammation, but no collection was noted. Active vasculitis with occluding thrombosis made us consider concomitant large vessel vasculitis like Takayasu arteritis along with Ankylosing spondylitis. Few reports of spondyloarthritis-associated vasculitis in the form of aortitis and peri aortitis have been described.

Because of active vasculitis, high disease activity of axial spondyloarthritis biologic DMARDs were planned to be initiated – anti-TNF inhibitors for targeting both musculoskeletal and vascular manifestations. As part of the safety workup, a Mantoux skin test was done which showed reactivity with 16mm induration. PET CT showed metabolic uptake at the transverse process of the C6 vertebra and anterior end of the right 2nd rib with slight surrounding inflammation, but no collection was noted. Hence a suspicion of Tuberculosis causing soft tissue inflammation/ phlegmon had arisen. After discussion with the Neurosurgery specialist and Radio diagnostician given the suspicion of Disseminated Tuberculosis with vertebral and vascular focus, a biopsy of lesions for histopathological diagnosis was planned.

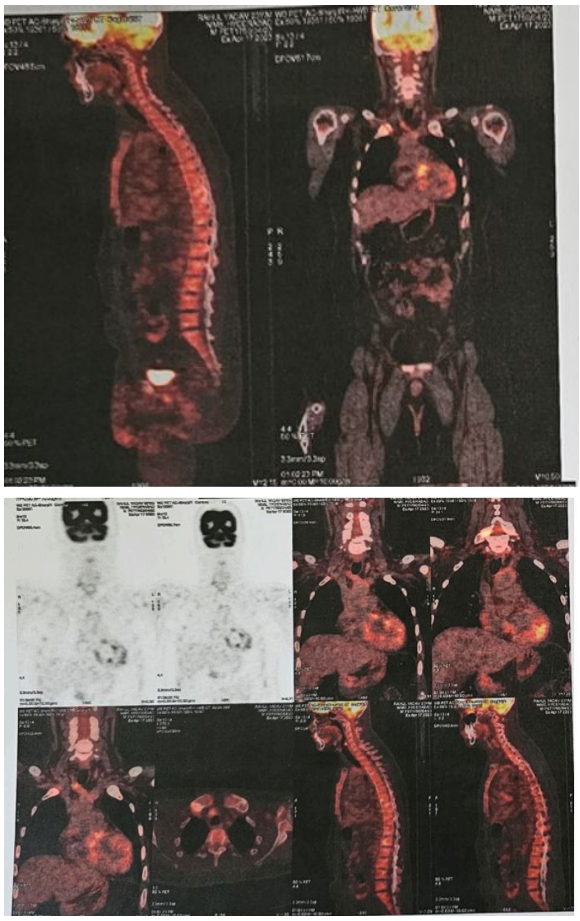

Figure 4: PET scan showing uptake in ascending, arch of aorta, and right 2nd rib

However, no definitive features of Tuberculosis were determined after reviewing PET-CT and CT angiography images. Anti-TNF agents were planned to be initiated given active sacroiliitis and active vasculitis, hence latent TB prophylaxis was initiated with two drugs (Isoniazid + Rifampicin).

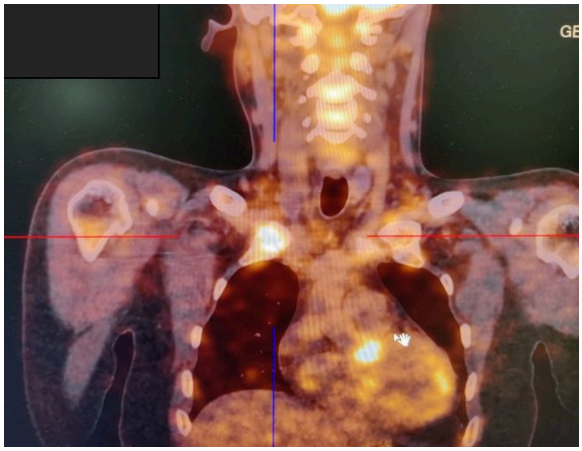

Figure 5: Metabolic uptake in the anterior end of right 2nd rib- probable pseudoarthrosis

Studies show that initiation of Anti TNF therapy with 3 weeks of Latent TB infection treatment is safe in Immune-mediated inflammatory diseases(1). Our patient presented for follow-up after 8 weeks of TB prophylaxis therapy, and Infliximab administered.

He had sudden onset weakness of the left lower face, left arm and leg and MR imaging of the brain revealed acute ischemic infarct in the right MCA territory involving parietal lobe. High-dose statin was added to the antiplatelet, and anticoagulation was added after 2 weeks of acute stroke.

Discussion

The prevalence of cardiovascular involvement in patients with Spondyloarthritis has been reported to be 10% to 30%. Aortic root involvement, valvular heart disease and conduction disturbances are the most common manifestations. Aortic involvement in Spondyloarthritis occurs during the late stages of the disease. Uncommonly aortic aneurysms can be seen in Spondyloarthritis and are usually accompanied by aortic insufficiency or aortic stenosis. Involvement of only the aorta sparing the valve is very rare. MA Khan observed that the prevalence of aortic valve disease was 4% in patients with a disease duration of less than 5 years and 10% in those with >30 years of disease. (2) Several reports have identified possible associations between vasculitis and autoimmune diseases, including spondyloarthropathies. (3)(4)

Association of Large vessel vasculitis like Takayasu arteritis and Ankylosing spondylitis is very rare. In a retrospective series analysis of 6 subjects done by Feng-ying Gan et.al., characteristically higher inflammatory markers, median ESR 84mm/hr, and median CRP 64mg/L were noted indicating a high inflammatory burden. (5) Duration between diagnosis of AS and Takayasu was 3-20 years, that is, they had longer disease duration. All subjects had peripheral joint involvement.

Due to reports of several cases of large vessel vasculitis along with Spondyloarthritis, a probable common etiopathogenic mechanism underlying both diseases has been postulated. Some authors believe the chronic underlying inflammation in Spondyloarthritis along with genetic predetermination, and molecular mimicry based on antigenic homologies between aorta and bone entheses might play a role in the association between these two rare disorders. (6)

In a retrospective survey of 14 patients in France presenting with SpA associated with Large vessel vasculitis - Takayasu arteritis, a major proportion of women, higher median age at SpA diagnosis, and greater frequency of HLA B27 negativity were noted. Elevated inflammatory markers were observed. (7) Recent studies show that IL12 can be one of the important inflammation mediators both in TA and SPA, polymorphisms of which have been observed in both disorders. Taken in context with the existing literature, it seems likely that an association between SpA with increased disease duration and large vessel vasculitides may exist. Findings suggest that chronic inflammation in axial SpA somehow triggers aortic inflammation.

In our case as well, long-standing uncontrolled spondyloarthritis could have triggered inflammation in extra musculoskeletal domains presenting as aortitis.

Conclusion

The presence of high inflammatory markers in Spondyloarthritis and long-standing uncontrolled disease activity should alert one to look for extra musculoskeletal manifestations in the form of vasculitis. Our patient highlights that Spondyloarthritis is a systemic disease with potentially severe extraarticular inflammation and consequences. If a patient presents with organ disease other than arthritis, it will be very difficult to meet diagnostic and classification criteria.

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