



A Case of Rheumatoid arthritis presenting with aorto-arteritis

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Abstract

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Introduction

Rheumatoid arthritis is a systemic inflammatory disease that has many extra-articular manifestations. Cardiovascular involvement, including coronary vasculitis and aortitis and skin lesion of erythema nodosum are uncommon findings of patients with rheumatoid arthritis, and thus, it is challenging for diagnosis of this case from those unusual extra-articular presentation. Though rare, a complex presentation of aortitis, erythema nodosum, and coronary complication can be found in an elder with rheumatoid arthritis. The patients with rheumatoid arthritis are often unrecognized to have associated cardiovascular disease.

Case Presentation:

A 54 year old female, came with complaints of breathlessness since 1 year, increased since 15 days (MMRC 2 aggravated to 3), cough with expectoration since 15 days, low grade fever since 2 days and multiple small joint pain associated with early morning stiffness since 15 years. There was no history of chest pain or hemoptysis.

The patient was admitted for further evaluation. Routine blood investigations were performed. All routine labs were within normal limit, whereas, RA factor came out to be positive 949.9 (0-30).

Chest radiography revealed mediastinal widening with apparent cardiomegaly.

High resolution computed tomography of thorax revealed Moderate circumferential wall thickening of aorta and the origin of brachiocephalic trunk and origin of left subclavian artery predominantly of the aortic arch with maximum thickness of 8.5mm suggestive of Aorto-arteritis. The right main pulmonary artery is significantly attenuated in caliber with more than 90% luminal compromise just posterior to aorta.

2 dimensional echocardiography revealed Moderate Tricuspid regurgitation with PASP of 47mmhg. There was no evidence of Pericardial effusion/clots/vegetations. Rheumatology reference was done and advised for USG Temporal artery, Tab Prednisolone 40mg OD, Tab MMF-S 360mg BD.

No abnormality was detected in USG Temporal artery.

Discussion:

Rheumatoid arthritis (RA) is a chronic systemic inflammatory disease primarily affecting synovial joints, but extra-articular manifestations are well recognized, particularly in long-standing, seropositive disease. Vasculitis in RA most commonly involves small- and medium-sized vessels; large-vessel involvement such as aorto-arteritis is exceedingly rare, making its recognition clinically significant.

Aortoarteritis in RA is thought to represent an immune-mediated process driven by persistent systemic inflammation. Proposed mechanisms include

immune complex deposition, endothelial injury, and cytokine-mediated vascular inflammation, particularly involving tumor necrosis factor- α and interleukin-6. Histopathological findings described in reported cases resemble those seen in other inflammatory large-vessel vasculitis, with granulomatous inflammation, lymphoplasmacytic infiltrates, and destruction of the elastic lamina.

Clinically, RA-associated aorto-arteritis may present insidiously with nonspecific symptoms such as fever, weight loss, fatigue, or elevated inflammatory markers, often preceding or overshadowing vascular symptoms. When symptomatic, patients may develop chest pain, back pain, limb claudication, pulse asymmetry, hypertension due to renal artery involvement, or complications such as aortic regurgitation and aneurysm formation. These features overlap with conditions such as Takayasu arteritis and giant cell arteritis, posing diagnostic challenges.

Imaging plays a pivotal role in diagnosis. Contrast-enhanced CT angiography and MR angiography typically demonstrate concentric aortic wall thickening, luminal narrowing, or aneurysmal dilatation. PET-CT may aid in identifying active vascular inflammation and assessing disease extent. In patients with established RA, especially those with high disease activity or seropositivity, the presence of unexplained systemic inflammation should prompt consideration of large-vessel vasculitis.



Conclusion:

There are no standardized treatment guidelines for RA-associated aorto-arteritis due to its rarity. Management is generally extrapolated from other inflammatory arteritis and includes high-dose systemic corticosteroids as first-line therapy. Conventional disease-modifying antirheumatic drugs (DMARDs) such as methotrexate, azathioprine, or cyclophosphamide may be used as steroid-sparing agents. Increasingly, biologic therapies targeting TNF- α or IL-6 have shown benefit in refractory cases, though evidence remains limited to case reports and small series. Surgical or endovascular intervention may be required in cases complicated by aneurysm, critical stenosis, or valvular involvement.

Early recognition of aorto-arteritis in RA is crucial, as delayed diagnosis may lead to life-threatening complications. This case underscores the importance of maintaining a high index of suspicion for large-vessel vasculitis in patients with RA presenting with systemic symptoms or unexplained inflammatory markers, even in the absence of classic vascular signs.

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