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Aortitis in a Patient with Psoriatic Arthritis

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Abstract: ***Aortitis refers to inflammation of the wall of aorta and includes both infectious and non - infectious etiologies. With the advances in imaging modalities and the increasing ease of availability, there is increasing incidence of aortitis and the phenotypic spectrum associated with the disease has widened. Giant cell arteritis and Takayasu arteritis, are the most common causes of non - infectious aortitis. Infectious aortitis can be a result of certain bacterial, viral or fungal pathogens, though uncommon, it can be devastating. Aortitis without a secondary cause or lack of involvement of other territories is defined as isolated aortitis. The management strategies and prognosis differs between aortitis subgroups which highlights the need for an extensive diagnostic workup. The areas of unmet clinical needs include proper disease classifications and lack of defined short - term and long - term management strategies. Here we report a case of aortitis in a middle aged lady who is a known case of psoriatic arthritis on treatment since past 5 years. We also address the diagnostic dilemmas in this case and discuss the available management strategies.***

1. Introduction

Primary vasculitic disorders account for less than 1% cases seen in a rheumatology clinic. Chandrasekaran et al and Samant et al reported an incidence of 0.38%, 0.44% respectively. Aortoarteritis accounts for 20% of the cases of vasculitis. (1) In a cohort of 1204 surgical aortic specimens studied by Rojo - Leyva et al, 168 (14%) had inflammation and 52 (4.3%) were classified as having idiopathic aortitis. (2) Vasculitis is common among women of reproductive age (female: male at a ratio of 9: 1) and is mostly detected at age 10 - 40 years. Vasculitis has a worldwide distribution, with the higher prevalence among Asians. Aortitis can present with constitutional symptoms, such as fever, weight loss, loss of appetite, or localized abdominal pain. The symptoms can be prolonged which can cause a delay in seeking medical attention. Inflammation of the aorta can result in aortic dilation, aortic insufficiency. Rarely, it can cause fibrous thickening of the aortic wall and ostial stenosis of its major branches, resulting in pulse and blood pressure discrepancy in the upper extremities. Hypertension could be secondary to renal artery stenosis. Depending on vessel involvement, claudication, neurological deficits, ocular disturbances, and other manifestations of vascular insufficiency can manifest in this disease.

Here we report a case of a 38 year old lady who is diagnosed with chronic plaque psoriasis with psoriatic arthritis and is on treatment for the same since the past 5 years. She has presented with subacute onset of left upper limb vascular insufficiency. Upon evaluation, she was found to have aortitis in PET CT and HLA B51 was positive on serological tests. We aim to highlight the diagnostic dilemmas we faced and elaborate various etiologies, diagnostic and management approaches for aortitis to emphasize the need for early detection and management.

2. Case Report

A 38 year old Asian women presented to Rheumatology OPD with discoloured digits of left upper limb since past two weeks which was associated with pain and paresthesias in her finger tips. The symptoms were persistent and progressive. She denied any precipitating factors. Notably, she was detected to have hypertension one month ago and was advised oral medications. Her past history was significant for chronic plaque psoriasis of ten years duration and polyarticular pattern of psoriatic arthritis of five years duration for which she was prescribed oral methotrexate. She was poorly compliant with the treatment in view of gastric intolerance and shifted to alternative medication for few years. On examination, there was cyanosis in left first, second, and fifth digits upto distal crease as observed in Fig 2. Skin examination showed psoriatic lesions over the scalp, extensor surfaces of elbows upto mid forearm, thighs, anterior surface of knees and legs, anterior abdomen and the entire back as observed in Fig 1. Nails and oral cavity examination was unremarkable and there was no lymphadenopathy. Heart rate was 90/min, blood pressure 130/90 mm Hg, Oxygen saturation of 98% on ambient air. There was no pulse or blood pressure discrepancy in the limbs but left radial arterial pulse was feeble. There was no carotidynia or carotid bruit. Systemic examination was unremarkable. There was no active arthritis or sacroiliitis.
The laboratory findings are summarized in table 1.

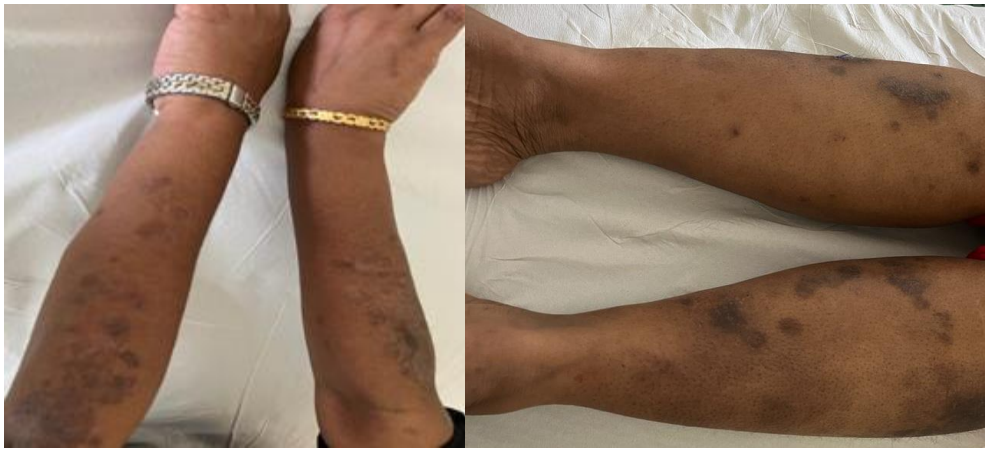

Figure 1: Extensive hypertrophied irregular lesions with raised margins over

Anterior aspect of legs, elbows and forearm, back and anterior abdominal wall

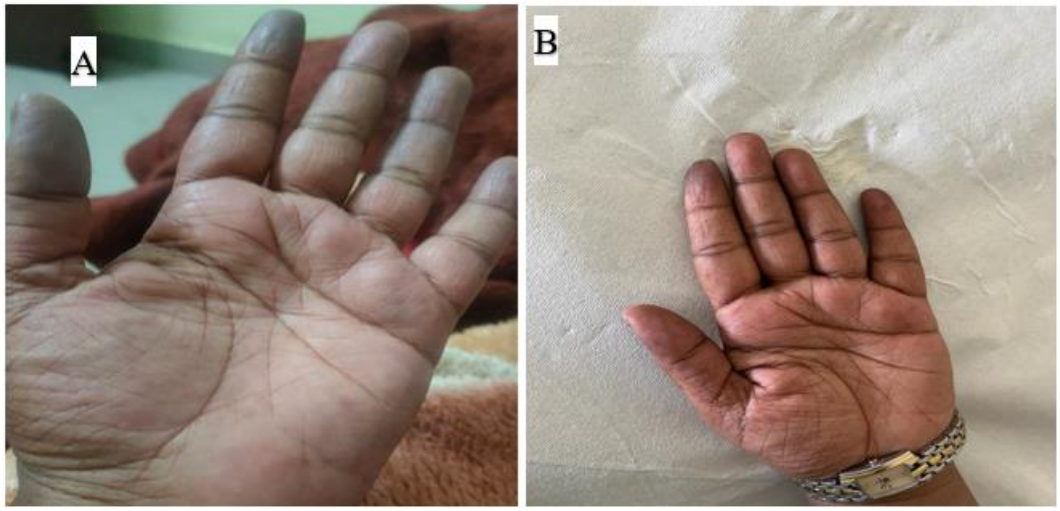

Figure 2: (A) Cyanosis over digital tips of left upper extremity upto the distal crease (B) post treatment improvement in cyanosis

HEMOGLOBIN	13.1g/dl
TLC	12100 cells/cumm
PLATELETS	3.7lakhs
LFT	Normal
RFT	Normal
ESR/CRP	6/NEG
VIRAL SEROLOGY	Negative
CUE	Normal
ANA IF	Negative
APLA PROFILE/ANCA	Negative
2DECHO	Normal
Left Upper Limb Doppler, CV Doppler	Normal Study
CT ANGIO OF Left Upper Limb Vessels	Normal

PET CT was done on day 6 of admission as a part of workup to rule out any large vessel vasculitis or malignancy. It showed metabolically active uptake in the walls of ascending, arch, descending thoracic aorta and abdominal aorta. Diffuse FDG uptake in both the parotid glands with fatty infiltration. There was no lymphadenopathy. A skin biopsy confirmed the diagnosis of psoriasis vulgaris. HLA B 51 was sent for vasculitis workup which was positive.

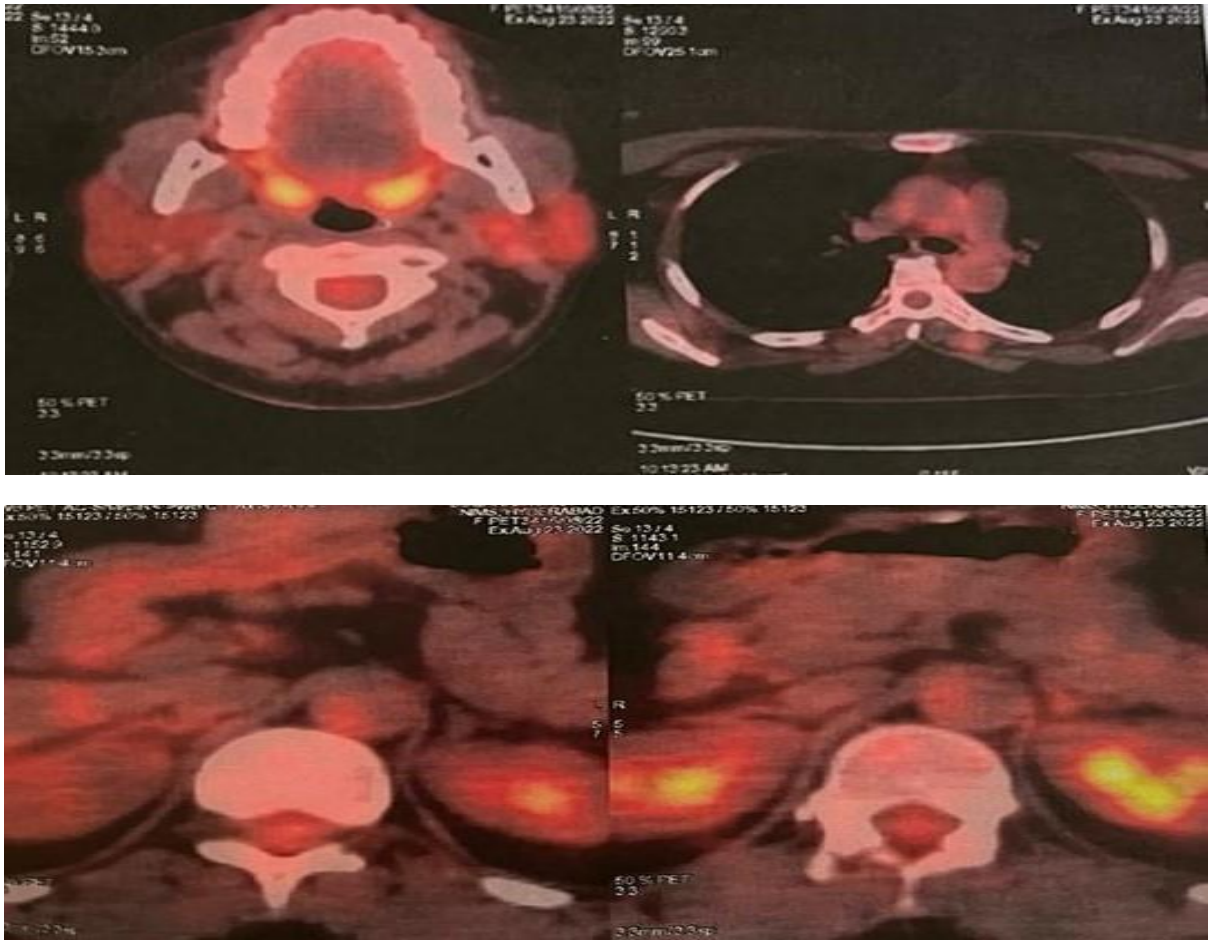

Figure 3: Metabolically active uptake in the walls of ascending, arch, descending thoracic aorta and abdominal aorta. Diffuse FDG uptake in both the parotid glands with fatty infiltration. No lymphadenopathy

She was managed with intravenous methyl prednisolone pulse therapy for 3 days and in view of active vasculitis initiated on induction immunosuppression with cyclophosphamide.6 months into the course of the disease, she had complete improvement of her symptoms with no further recurrence so far. She is currently on maintenance immunosuppression with methotrexate 20mg weekly along with a low dose oral glucocorticoid.

3. Discussion

Aortitis can be a result of either infectious or non - infectious etiologies. Bacteria such as Streptococcus pneumonia, Streptococcus pyogenes, Staphylococcus, and Salmonella species are the most prevalent infectious causes. Rheumatological diseases are the most common non - infectious causes. Among the Rheumatological causes, primary large vessel vasculitis which includes Giant cell arteritis and Takayasu arteritis are the most common causes of aortitis. Aortitis can also be seen in other rheumatological diseases with a lower incidence in Systemic lupus erythematosus, Rheumatoid arthritis, HLA B27 associated arthropathies like long standing Ankylosing spondylitis and Reiters disease, Behcet's disease, Cogans, Sarcoidosis and IgG4 disease.

Aortitis develops in three phases. Phase I is the inflammatory period presenting with nonspecific systemic symptoms which may include low - grade fever, weight loss, fatigue, arthralgia, and elevated inflammation markers (Erythrocyte sedimentation rate, C - reactive protein) without any loss of peripheral pulses. Phase II involves inflammation of the vessel wall which is associated with pain (Carotidynia) and/or tenderness of the arteries. Phase III is characterised by permanent wall injury, which can present with signs and symptoms of vascular insufficiency secondary to narrowing, or occlusion, dilation, intramural tearing of the proximal or distal branches of the aorta. In advanced cases, occlusion of the vessels of the extremities may cause ischemic skin changes, ulcerations or gangrene.

Our patient had a subacute onset of aortitis with HLA B51 positivity but lacked the typical features of Behcet disease like orogenital ulcers, ocular manifestations or skin lesions. HLA B51 positivity alone cannot substantiate the etiology of aortitis in this scenario. The risk of *HLA–B51/B5* carriers developing BD is increased by a factor of 5.78. Rates of *HLA–B51/B5*–positive BD cases varies across geographic locations. Though there is a clear association with the disease, this genetic association might hold true only for areas of high BD prevalence. (3) Vascular complications in Bechet occur in 14% of patients with 2% presenting with vascular symptoms at onset. Most common vascular symptoms are superficial vein thrombosis followed by DVT and arterial complications as presenting features are rare. According to few case reports, Behçet disease may place an individual at a significantly increased risk of psoriasis, and still greater hazard of being affected with psoriatic arthritis. In age - group sub - analysis, individuals over 65 years with skin psoriasis were one and a half times more likely to be affected with Behcet disease,

relative to those under 65. Moderately obese (30–35 kg/m^2) " group have an Odds ratio of 1.24 (95% CI 1.12–1.38, $p < 0.001$) to be associated with Psoriatic Arthritis (4) . Our patients demographics did not satisfy these associations though she tested positive for HLA B51

With the lack of other features of CTD, SLE and RA were not considered as our differential diagnosis. Involvement of the aorta is well defined in HLA - B27 - associated spondyloarthropathies such as long - standing ankylosing spondylitis and Reiter's syndrome. However, true aortitis is not a typical feature of psoriatic arthritis and has been reported in only a few cases (5) . Sarcoidosis and Cogans were also not considered as there were no supporting features specific for the diseases. IgG4 related disorder was another possible differential diagnosis as PET CT showed uptake in both parotid glands. Typical features of IgG4RD like lymphadenopathy, organomegaly were absent and patient denied any parotid swelling or pain which was confirmed on examination. Hence, IgG4RD was excluded as a cause of aortitis.

Psoriatic arthritis is a rare cause of aortitis and is described in few case reports wherein it is associated with a long standing, extensive psoriasis and destructive psoriatic arthritis. Our patient similarly had extensive skin psoriasis at the time of onset of aortitis and was not receiving any medications. Interestingly, her symptoms of aortitis and skin psoriasis improved drastically with cyclophosphamide. She was continued on oral Leflunomide for psoriatic arthritis as she had intolerance to methotrexate. After 3 months of follow up, her skin psoriasis and psoriatic arthritis is in remission and she reports no further symptoms of vascular insufficiency.

The diagnosis of aortitis is made on the basis of clinical presentation and aortic imaging. The diagnostic workup of a patient with suspected aortitis should include erythrocyte sedimentation rate (ESR) and c - reactive protein (CRP), a complete blood count, assessment of kidney and liver function, and blood cultures to exclude infectious aortitis.

Additional laboratory testing should be based upon the clinical assessment of the patient and the differential diagnosis considered. Anti - nuclear antibodies, anti - neutrophil cytoplasmic antibodies, and rheumatoid factor may be helpful in the appropriate clinical setting. Skin testing for tuberculosis and serologic testing for syphilis can be done when clinical suspicion of these disorders is high.

Once the diagnosis of aortitis has been made, the approach to management depends upon the etiology. The goals of therapy include immediate treatment of aortic inflammation along with maintenance therapy and management of arterial complications.

4. Conclusion

Psoriatic arthritis, though a rare cause of aortitis, must be considered in patients with long standing and extensive psoriasis particularly in untreated patients when they present with signs and symptoms of vascular insufficiency. A multidisciplinary approach is advocated in diagnosing and managing a patient with aortitis. Non - infectious aortitis which is the most common cause requires extensive workup to identify the underlying etiology. A high index of suspicion is necessary in these disorders, as it is important to advocate early immunosuppression to avoid dreaded complications.

Conflict of Interest: None of the authors have any conflicts of interest.

Patients written informed consent has been taken

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