

Thoracic peri-aortic fibrosis in a patient of psoriasis – Cyclosporine as a putative etiologic agent

Nitin P Ghonge^{1*}, Rajesh Gothi Gothi¹

1. Diwanchand Imaging and Research Centre New Delhi, India

* **Correspondence:** Dr. Nitin P. Ghonge, Diwanchand Imaging and Research Centre, 10 B, Kasturbha Gandhi Marg, New Delhi-110001, India

 drnitinhonge@rediffmail.com :: Tel: 91-09868452890

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ABSTRACT

The article describes an unusual occurrence of peri-aortic fibrosis with consequent luminal stenosis in descending thoracic aorta in an adult case of Psoriasis. The report also illustrates the role of Multi-detector CT in the diagnosis of thoracic peri-aortic fibrosis. The patient had received cyclosporine on multiple occasions during acute exacerbation of disease. In absence of any concomitant infective-inflammatory system disorder or atherosclerotic process, the cyclosporine is suggested as a putative etiologic agent for peri-aortic fibrosis.

CASE REPORT

Introduction

Peri-aortic fibrosis is a well-established entity which is described in relation with variety of etiologic causes. Atherosclerosis, infective-inflammatory diseases like rheumatology conditions and tuberculosis are often associated with peri-aortic fibrosis. We present a unique case in that it was devoid of any underlying systemic cause.

Our patient had received cyclosporine on multiple occasions during acute exacerbation of disease for psoriasis. In absence of any concomitant infective-inflammatory system disorder or atherosclerotic process, the cyclosporine is suggested as a putative etiologic agent for peri-aortic fibrosis.

Case report

A 35 year-old man presented to our institution with complaints of intermittent claudication in both lower limbs. He denied pain at rest. A review of his medical systems showed that the patient had long-standing history of Psoriasis with multiple episodes of remissions and recurrences. He had received Cyclosporine on multiple occasions during the acute phase of his disease. On clinical examination, there were multiple erythematous skin lesions with scaling in the trunk and extremities (Figure 1). The patient was also found to have hypertension with brachial arterial pressure readings of

160/120 mm of Hg and popliteal arterial readings of 110/70. Multi-detector CT (MDCT) Angiography showed a circumferential smooth soft tissue thickening encasing the distal part of descending thoracic aorta causing significant luminal stenosis (Figure 2). The aortic wall was visualized separate from the periaortic soft tissue as a thin hypodense rim, between the contrast-filled lumen and the soft tissue (Figure 3). The peri-aortic soft tissue is not extending up to the diaphragmatic hiatus (seen in the interactive mode). The abdominal aorta, iliac, femoral, popliteal and the distal leg arteries do not show any definite abnormality. Biopsy was not performed to establish the diagnosis, as the patient did not give consent for the same. The CT images were primarily studied by two radiologists (NPG and RG) and a consensus-based diagnosis of thoracic peri-aortic fibrosis was made in the clinical-radiological meeting of the institute. The possibilities of atherosclerotic or Takayasu's disease were considered unlikely. The key finding against these conditions was the demonstration of hypodense aortic wall between the contrast-filled lumen and the peri-aortic soft tissue. In addition, there was no vascular lesion elsewhere. In absence of any other concomitant infective-inflammatory cause, the cyclosporine was accepted as a likely causative factor for the peri-aortic disease process.

The patient underwent an endovascular dilatation procedure with stent placement in the same sitting. The procedure was uneventful without any significant post-

procedural complications. Patient reported marked improvement in the symptoms of intermittent claudication in routine follow-up examination at sixth months and twelfth months. The brachial arterial pressure readings were 130/90 mm of Hg and popliteal arterial readings were 124/80, one year after the endovascular treatment.

DISCUSSION

An extensive review of the medical literature includes case reports of thoracic periaortic fibrosis with associated infective-inflammatory diseases such as rheumatology conditions and tuberculosis or had involvement of the abdominal aorta (1). Inflammatory aneurysm is an aortic aneurysm disease characterized by significant peri-aneurysmal inflammation and adhesions to surrounding structures (2). The inflammatory aneurysms are thought to be related to peri-aortic retroperitoneal fibrosis and various autoimmune diseases, including rheumatoid arthritis, systemic lupus erythematosus, and giant cell arteritis (3).

A recent article also described a similar case with pathologically-confirmed isolated thoracic peri-aortic fibrosis in a young male who presented with back pain. In this case, there was intense accumulation of Fluoro-deoxy-glucose on Positron emission tomography (FDG-PET) (4). Segers B et al described a young male with Takayasu's disease who had an isolated stenosing lesion in the lower thoracic aorta and presented with upper extremity arterial hypertension, weak femoral pulses and a systolic bruit in lower inter-scapular region (5). Though, our case bear superficial resemblance to the case reported by Segers B et al, the aortic stenosis in our case is more 'focal' as compared to long-segment stenosis in the above-mentioned case. Moreover, unlike our patient, no mention is made about the separate visualization of the aortic wall in the stenotic segment in this case. The key finding against Takayasu's arteritis in our patient was the demonstration of hypodense aortic wall between the contrast-filled lumen and the peri-aortic soft tissue (black arrowhead in figure 3). Takayasu's disease is essentially a condition involving preferentially the aortic/arterial wall rather than the peri-aortic soft tissue. In addition, there was no vascular lesion elsewhere. Middle aortic syndrome was the term coined by Sen et al to describe inflammatory sub-isthmic aortic narrowing (6). Inflammatory diseases such as Takayasu's arteritis, aorto-arteritis and granulomatous arteritis are described to be common causes of sub-isthmic stenoses (7). Our patient however, did not have any underlying infective-inflammatory mediastinal disease, peri-aortic disease in the abdomen or any other vascular, auto-immune or connective tissue disorder. There are reports of inflammatory reactions to endo-luminal stents that itself lead to peri-vascular thickening and peri-aortitis (8, 9). This also emphasizes the need for long-term follow-up in our patient.

In our case, there is isolated thoracic periaortic fibrosis in a patient with long-standing history of Psoriasis, who had received Cyclosporine at multiple occasions during the acute phase of disease. Cyclosporine-induced fibrogenic effects are previously described in animal studies (10, 11). The present case report is unique in terms of putative etiologic role of cyclosporine, which is not previously described. Interestingly,

the cyclosporine is also shown to cause remission in patients with retroperitoneal fibrosis (12). The exact cause-effect relationship warrants further investigation with appropriate clinical trials.

TEACHING POINT

Cyclosporine might be a causative agent for peri-aortic fibrosis. Multi-detector CT plays a vital role in the diagnosis of thoracic peri-aortic fibrosis.

ABBREVIATIONS

MDCT = Multi-detector CT

FDG-PET = Fluoro-deoxy-glucose Positron emission tomography

REFERENCES

1. Jois RN, Gaffney K, Marshall T and Scott DGI. Chronic periaortitis Rheumatology 2004 43(11):1441-1446.
2. Walker DI, Bloor K, Williams G, et al. Inflammatory aneurysms of the abdominal aorta. Br J Surg 1972;59:609-614.
3. Haug ES, Skomsvoll JF, Jacobsen G, et al. Inflammatory aortic aneurysm is associated with increased incidence of autoimmune disease. J Vasc Surg 2003;38:492-497.
4. Lee YK, Seo JB, Kim SS and Lim TH. Case Report : Thoracic Periaortic Fibrosis Mimicking Malignant Tumor: CT and 18F-FDG-PET Findings. AJR 2007; 188:345-347.
5. Segers B, Derluyn M, Barroy JP, Brunet AP. Isolated supradiaphragmatic descending thoracic aorta stenosis in a Takayasu's disease: surgical cure Eur J Cardiothorac Surg 2001;20:1243-1245.
6. Sen P.K., Kinare S.G., Engineer S.D., Parulkar G.B. The middle aortic syndrome. Br Heart J 1963;25:610-618.
7. Watson N.A., Chalmers N., Navqui N. Suprardiaphragmatic middle aorta syndrome – MR and angiographic imaging. Br J Radiol 1998;71:213-216.
8. Schürmann K, Vorwerk D, Bücken A, et al. Perigraft inflammation due to Dacron-covered stent-grafts in sheep iliac arteries: correlation of MR imaging and histopathologic findings. Radiology 1997;204:757-763.
9. Simons PG, Overhagen HV, Bruijninx CA, Kropman RF and Kuijpers KC. Periaortitis with Ureteral Obstruction After Endovascular Repair of an Abdominal Aortic Aneurysm. AJR 2002; 179:118-120.
10. Rita R; Luigi R; Barbara B; Lorena G; Alessandra S ; Rossella B. Cyclosporine A induces vascular fibrosis & heat shock protein expression in rat. International Immunopharmacology 2005, vol. 5; 69-176.
11. Yang G, Meguro T, Hong C, Asai K, Takagi G, Karoor VL, Sadoshima J, Vatner DE, Bishop SP, Vatner SF. Cyclosporine reduces left ventricular mass with chronic aortic banding in mice, which could be due to apoptosis and fibrosis. J Mol Cell Cardiol. 2001 Aug;33(8):1505-14.
12. Marzano A, Trapani A, Leone N, Actis GC, Rizzetto M. Treatment of idiopathic retroperitoneal fibrosis using cyclosporine. Ann Rheum Dis 2001;60:427-428.

FIGURES

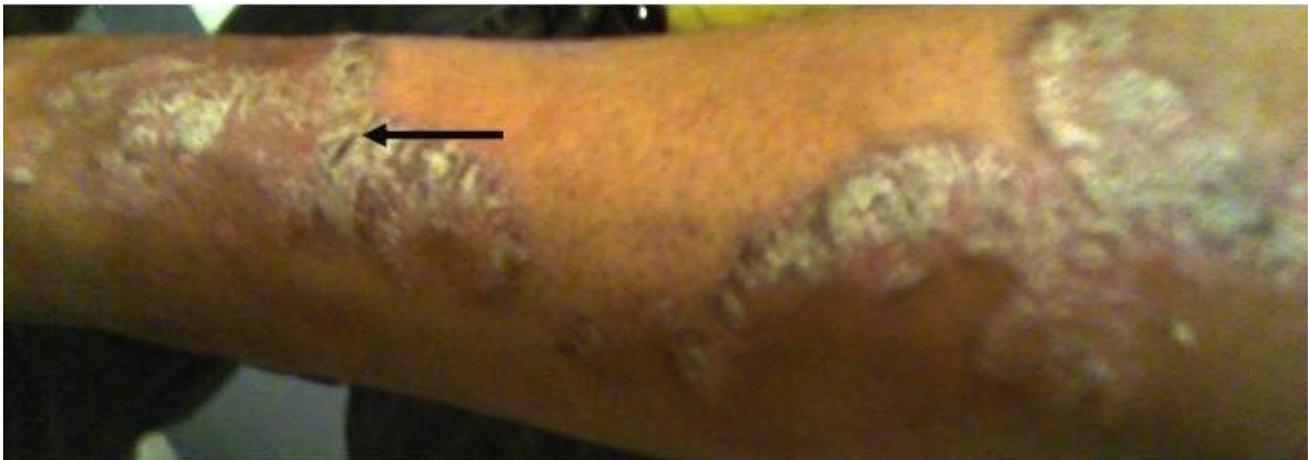


Figure 1: Clinical photograph of the patient’s left upper limb showing extensive psoriatic disease with areas of erythema and scaling (arrow).

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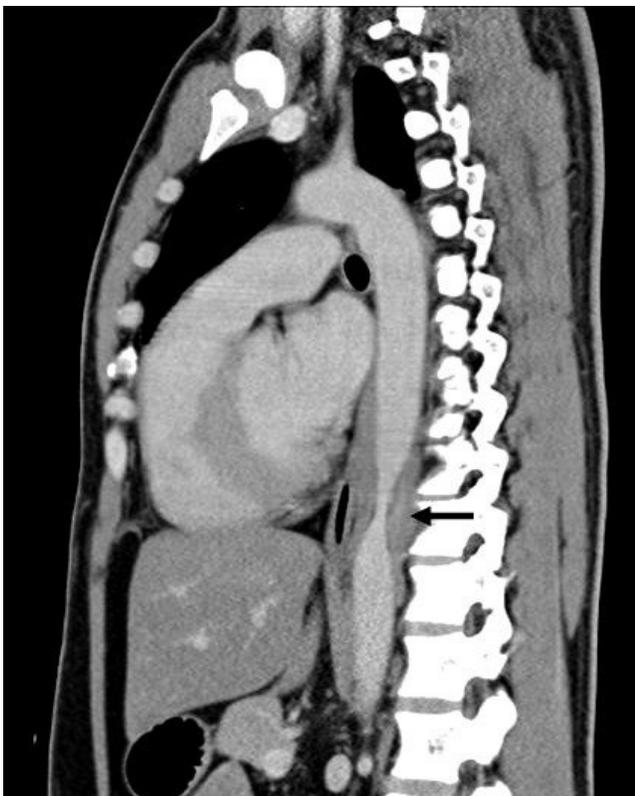


Figure 2: Sagittal multiplanar reformatted contrast enhanced multi-detector CT (MDCT) image showing periaortic soft tissue causing significant luminal stenosis in the distal thoracic aorta (arrow).

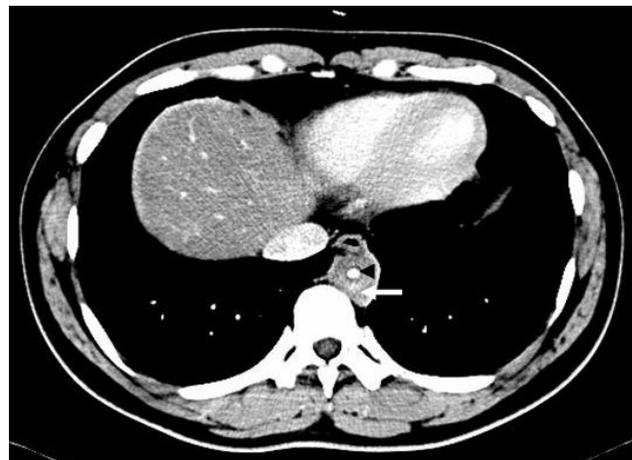


Figure 3: Transverse contrast enhanced multi-detector CT (MDCT) image showing circumferential distribution of the periaortic soft tissue (white arrow). The aortic wall can be separately visualized (black arrowhead).

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