Takayasu Arteritis with Atrial Septal Defect Presenting as Sterile Corneal Melt

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Abstract

Sterile corneal melt in a previously healthy 40 year female with features of Takayasu arteritis with incidental atrial septal defect is presented.

Introduction

Takayasu arteritis is a granulomatous vasculitis of large vessels with a predilection of aorta and its main branches and pulmonary arteries. Patients usually present with claudication, headache, syncope or constitutional symptoms like fever, malaise, night sweats. The ocular manifestations are not uncommon in Takayasu arteritis. These include ocular ischemic syndrome, hypertensive retinopathy, Takayasu retinopathy.[1,2] Sterile corneal melt is a rare presentation of Takayasu arteritis.

This is a case of 40 year female who presented with decreased vision in the right eye, incidentally found to have absent pulses and low blood pressure in both upper limbs. Her CT angiogram findings were suggestive of Takayasu arteritis (Figures 1, 2). So this article implicates a rare presentation of Takaysu arteritis.

Case Profile

A 40 year female initially admitted in ophthalmology ward with history of decreased vision in the right eye for past 1 month, history of pain and redness in the right eye one month back. Ocular examination revealed a sterile corneal melt with iris prolapse in the right eye (Figure 4). She was found to have very low blood pressure and absent pulses in both upper limbs. Patient was referred for physician opinion.

The patient is totally asymptomatic other than ocular symptoms. General examination was normal. On examination of the peripheral pulses, the brachial, radial and ulnar pulses were absent on both upper limbs. She had a carotid thrill with bruit on the right side. The blood pressure in right upper limb was 80/50 mmHg, left upper limb 70/50 mmHg, right lower limb 140/80 mmHg and left lower limb was 150/70 mmHg. There was a wide and fixed second heart sound with ejection systolic murmur of grade 2/6 in pulmonary area. Her respiratory system, abdomen, central nervous system were normal. Fundus was also normal.

Her blood investigations revealed elevated ESR 100mm/hr, positive CRP. RF, HBsAg, VCTC, VDRL, Mantoux were negative. Her renal and liver function tests were normal. ECG showed features of right axis deviation with features RV overload. Chest X-Ray was normal. Echocardiography showed dilated Right atrium and ventricle with Atrial septal defect of ostium secondum type left to right shunt with moderate PAH (Figure 3).

Arterial Doppler of both upper limbs showed diffuse wall thickening of both subclavian arteries with high velocity biphasic flow and a low velocity monophasic flow in both axillary and brachial arteries. There was a venous like flow with no pulsatility in both radial and ulnar arteries. There was no evidence of thrombosis.

Carotid artery Doppler showed diffuse intimal and medial thickness of common carotid and internal carotid arteries on both sides with increased flow velocity. There was no evidence of plaque.

CT Angiogram

Ascending aorta (2.8 cm), arch of aorta (2.6 cm) and descending aorta (1.9cm) were normal in callibre. Branches of aortic arch like brachiocephalic artery, left CCA and left subclavian artery showed severe ostial stenosis with severe narrowing right and left CCA and subclavian artery (Figures 1, 2). Both vertebral artery were smaller in callibre. These features were suggestive of Takaysu Artertis Type 1.

She was treated with injectable steroids and tectonic keratoplasty was done in the right eye (Figure 5). The vision in the right eye was improved in the postoperative period. The inflammatory markers ESR, CRP has come down with the treatment. She was on oral prednisolone 1mg/kg/day.

Discussion

Takayasu arteritis is an uncommon disease with an incidence of 1.2-2.6 / million/year. It is more prevalent in women of child bearing age. 10-20% are clinically symptomatic. ACR 1990 classification criteria for Takayasu arteritis includes

1. Age at onset ≤ 40 years
2. Claudication of extremities
3. Decreased/ absent brachial artery pulsation
4. Blood pressure of > 10mmHg difference between the extremities
5. Bruit over subclavian arteries or carotid
6. Arteriogram abnormality

3 out 6 criteria is needed for the diagnosis. Our patient had 5 out of these 6 criteria except for the claudication.

Takayasu arteritis can be classified into 6 types based on the angiographic involvement.

Type 1: Branches of the aortic arch

Type 2A: Ascending aorta, aortic arch and its branches

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Fig. 1: CT Angiogram showing stenosed left common carotid artery. The aortic arch is normal in callibre

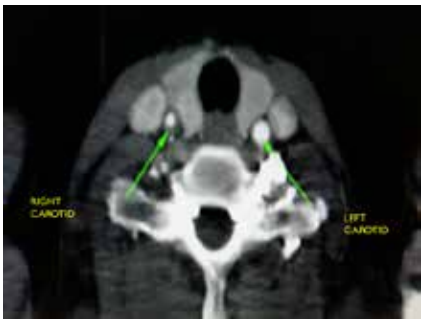

Fig. 2: CT angiogram showing the small callibre of both carotids

Type 2B: Type 2A region plus thoracic descending aorta

Type 3: Thoracic descending aorta, abdominal aorta, renal arteries or its combination.

Type 4: Abdominal aorta, renal arteries or both

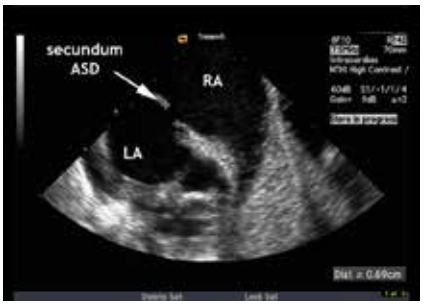

Fig. 3: Echocardiography showing Atrial septal defect

Fig. 4: Sterile corneal melt in the right eye

Type 5: Entire aorta and its branches

In India, abdominal aorta is the most common vessel to be involved.[3] In our patient only the branches of aortic arch (Type 1) were involved.

Peripheral corneal ulceration or corneal melting is not infrequent

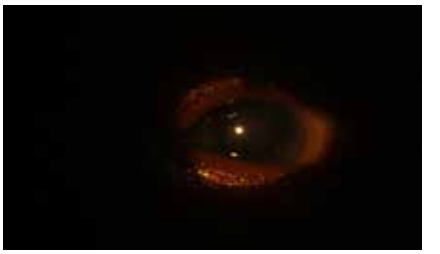

Fig. 5: Post keratoplasty picture of the right eye

occurrence in patients with systemic diseases like Rheumatoid arthritis, SLE, Polyarteritis Nodosa and Giant cell arteritis. Takayasu arteritis presenting as sterile corneal melt has not been reported so far anywhere.[4] The presence of ostium secondum ASD in our patient was possibly coincidental.

In conclusion, our report not only presents an unusual ocular presentation of Takayasu arteritis, it also reiterates the importance of systemic examination in a patient with ocular presentation.

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