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Case report

Atypical Tolosa Hunt syndrome with bilateral sclerokeratitis and optic atrophy in Takayasu's arteritis

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ABSTRACT

Purpose: We report an interesting case of atypical Tolosa Hunt syndrome with bilateral Sclerokeratitis and optic atrophy in a patient with Takayasu's arteritis.
Observations: A 31-year- old lady presented with severe retroorbital pain in the right eye and right sided headache of 2 weeks duration. She had a history of 6th and 7th nerve palsies which improved with oral corticosteroids. Her BCVA was no perception of light in OD and 6/6 in OS. Slitlamp examination showed bilateral old sclerokeratitis and fundus examination showed bilateral optic atrophy. Systemic examination revealed BP of 240/100 mm of hg in the right upper limb with absent pulsations in the left radial, bilateral femoral and dorsalis pedis arteries noted. CT aortic angiography confirmed the diagnosis of Takayasu's arteritis. Symptoms improved with oral corticosteroids and azathioprine, which was followed by a renal stenting procedure. At 1 year followup, she is doing well with no recurrences.
Conclusions and importance: This case report presents a unique manifestation of atypical Tolosa Hunt syndrome in Takayasu's arteritis. To our knowledge, atypical Tolosa Hunt syndrome in Takayasu'arteritis has not been described in literature.

1. Introduction

We report an interesting case of atypical Tolosa Hunt syndrome with bilateral Sclerokeratitis and optic atrophy in a patient with Takayasu's arteritis. To our knowledge this association in Takayasu's arteritis has not been described in literature (Medline search).

2. Case report

A 31-year- old lady was referred with a history of severe retroorbital pain in the right eye and right sided headache of 2 weeks duration. She also gave a history of redness, pain and decrease in vision in both eyes since 3 years for which she had received oral corticosteroids on and off elsewhere. Systemic history was significant for a right sided 6th and 7th cranial nerve palsies 2 years ago which recovered with oral corticosteroids. She had no history of diabetes mellitus. On examination, her BCVA was no perception of light and 6/6 in OD and OS respectively. Relative afferent pupillary defect was noted in OD. Slit lamp examination showed areas of old scleritis with peripheral corneal involvement suggestive of old sclerokeratitis (Fig. 1) Fundus examination revealed bilateral optic atrophy (Fig. 2A&B). Systemic examination revealed BP of 240/100 mm of hg in the right upper limb with absent pulsations in the left radial, bilateral femoral and dorsalis pedis arteries noted. Investigations showed an ESR of 60 mm/hr and high serum homocysteine levels. Rheumatoid factor, antinuclear antibody, antineutrophilic cytoplasmic antibody, serum angiotensin converting enzyme, mantoux test were negative. Random blood sugar, routine urine and serum creatinine were normal. MRI orbit and brain showed an inflammatory mass lesion in the right orbital apex(Fig. 3). CT aortic angiography revealed features of aortoarteritis, mild fusiform aneurysm of ascending aorta and proximal aortic arch. There was diffuse wall thickening with multiple foci of narrowing involving aortic arch, descending thoracic aorta, abdominal aorta, left subclavian artery, right external iliac artery and occlusion of proximal celiac artery(Fig. 4). A diagnosis of Takayasu's arteritis with atypical Tolosa Hunt syndrome, old sclerokeratitis and optic atrophy was made. She was started on oral corticosteroids (1mg/kg body weight) and azathioprine (eventually increased upto 1.5mg/kg bodyweight). There was a marked

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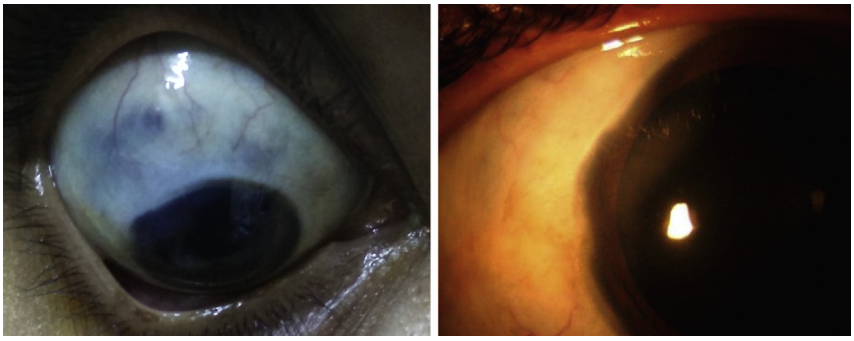

Fig. 1. External photograph showing scleral thinning and peripheral corneal scar.

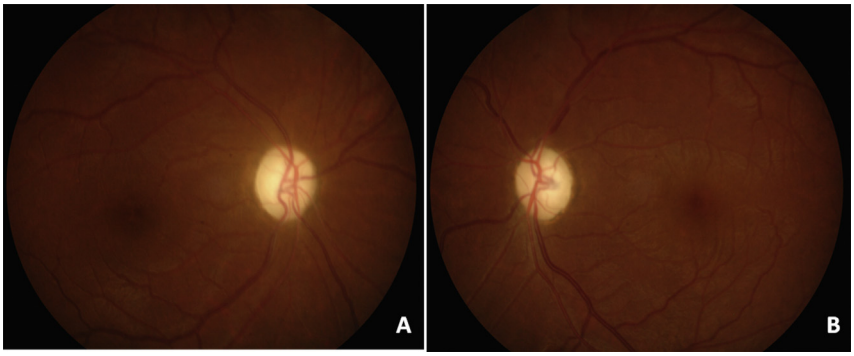

Fig. 2. Fundus photograph showing optic atrophy in OD(A) and OS (B).

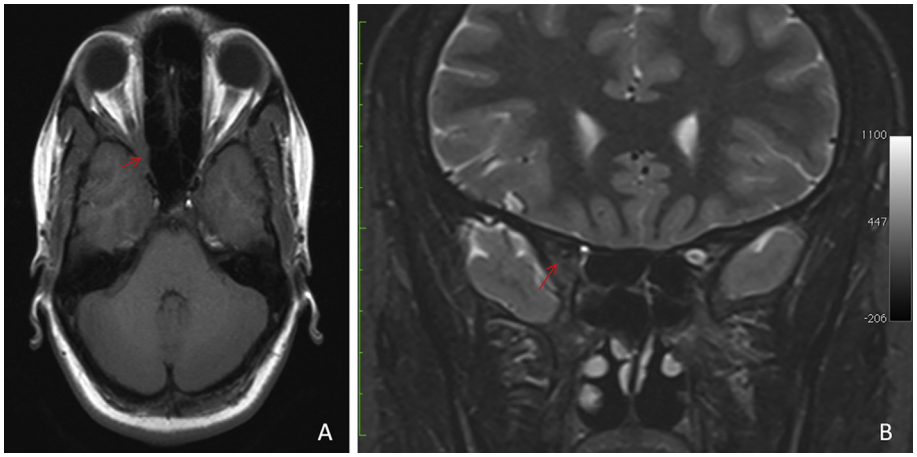

Fig. 3. Magnetic Resonance Imaging of orbit and cranium showing axial T1 weighted image showing soft tissue mass appearing hypo intense at right orbital apex compressing right optic nerve (A) and coronal stir image showing soft tissue mass appearing minimally hyper intense at right orbital apex compressing the right optic nerve (B).

improvement in pain, She eventually underwent stent placement for the renal artery stenosis. At 1 year followup, she is doing well with no ocular pain.

3. Discussion

Retinopathy, vascular occlusions, ocular ischaemic syndrome, anterior ischaemic optic neuropathy, scleritis[1] and recently pseudotumor[2] have been reported in Takayasu's arteritis. Our case of Takayasu's arteritis is unique in having an inflammatory mass in the right orbital apex and a history of right 6th and 7th cranial nerve palsies in the past, which resolved with oral corticosteroids, suggestive of an atypical Tolosa Hunt syndrome. Facial nerve palsy though atypical, has been reported in Tolosa Hunt syndrome[3] and is suggestive of a generalised inflammatory process.[4] Ideally a biopsy of the inflammatory lesion would have helped to further establish the diagnosis. However in view of the systemic condition and dramatic improvement with oral corticosteroids, the potential risks with such a biopsy was unacceptable.

4. Conclusion

This case highlights a unique presentation of atypical Tolosa Hunt syndrome with bilateral sclerokeratitis and optic atrophy in a patient with Takayasu's arteritis.

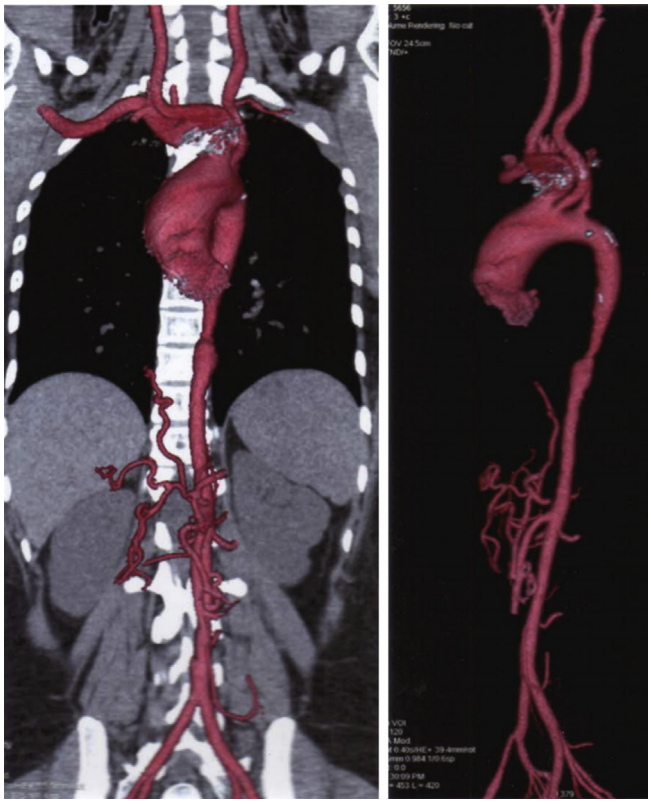

Fig. 4. Computed tomography of aortic angiography showing features of aorta arteritis with multiple foci of narrowing involving left subclavian artery, aortic arch and descending aorta.

Patient consent

Institutional review board approval and written patient consent to publish case details such as photographs in the case report have been obtained.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Conflict of interest

The authors have no financial disclosures.

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