

## Letter to the Editor (Case report)

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### Recurrent scleritis as a presenting manifestation of asymptomatic occult Takayasu arteritis

#### Key message

- Takayasu arteritis should be in the list of differential diagnoses in patients with isolated scleritis.

DEAR EDITOR, Takayasu arteritis is a rare chronic granulomatous large-vessel vasculitis with preferential involvement of the aorta, its major branches and the pulmonary arteries [1]. Ocular involvement in Takayasu arteritis is seen as Takayasu retinopathy, which is the result of ocular hypoperfusion and chronic ischaemia [2]. Scleritis in Takayasu arteritis is extremely rare, reported in only six cases [3–6]. We report a case of previously undiagnosed asymptomatic Takayasu arteritis presenting with bilateral recurrent anterior scleritis.

A 48-year-old woman presented with redness and pain in the right eye for 15 days. Pain radiated around the eye and was exacerbated by eye movements. She had history of multiple episodes of inflamed, painful eyes for past 6 years, involving the left eye for the initial 4 years and the right eye thereafter. She received topical CSs and NSAIDs, and oral CSs at variable doses during these episodes.

Examination of the eyes showed temporal congestion in the right eye (Fig. 1A) and a normal left eye. Ophthalmic assessment confirmed nodular anterior scleritis of the right eye. Visual acuity, the cornea, anterior and posterior chambers were normal in both eyes. Further physical examination revealed absent pulses in the left radial, ulnar, brachial and subclavian arteries. Right radial and brachial pulses were diminished. Bruits were heard over bilateral carotid and left subclavian arteries. Blood pressure was not recordable in the left arm, 90/60 mmHg in the right arm and 130/80 mmHg in both lower limbs.

On evaluation, ESR was 49 mm/h, CRP was 20.4 mg/l, and ANCA (ELISA and immunofluorescence) and ANA tests were negative. Chest radiographs and echocardiographs were normal. CT angiogram showed circumferential wall thickening in the ascending (arrow in Fig. 1B), arch and descending thoracic aorta (arrowhead in Fig. 1B) and the brachiocephalic, left common carotid and left subclavian arteries. Complete occlusion was seen in the proximal part of the left subclavian artery (arrow in Fig. 1C), with collaterals filling the distal segment.

Stenosis was seen in the right common carotid and bilateral subclavian arteries (arrowheads in Fig. 1C). A diagnosis of Takayasu arteritis with anterior nodular scleritis was made, and oral prednisolone was started at 1 mg/kg in combination with oral MTX 15 mg/week. Ocular symptoms improved markedly over the next few days, and gradual tapering of prednisolone dose was planned.

We have found only six published cases of Takayasu arteritis associated with scleritis [3–8]. Akhtar *et al.* [3] reported a case with a 10-year history of Takayasu arteritis. The patient was in prolonged remission before developing scleritis as a presenting manifestation of disease flare. Scleritis was refractory to MMF and required adalimumab for CS weaning. Scleritis as a presenting manifestation in an asymptomatic occult Takayasu arteritis was reported only once [7]. This patient succumbed to ischaemic colitis 3 weeks after presentation, and necrotizing granulomatous vasculitis of the thoracic and abdominal aorta was demonstrated on autopsy. Similar to our case, Chaudhary *et al.* [8] also reported a case of Takayasu arteritis with a 6-year-long history of fluctuating scleritis. But unlike their patient, our patient was asymptomatic for Takayasu arteritis and had occult vascular inflammation for an unknown duration, leading to stenosis and occlusion of vessels. This presentation makes our case extremely unusual and also emphasizes the importance of detailed physical examination even in patients presenting with isolated scleritis.

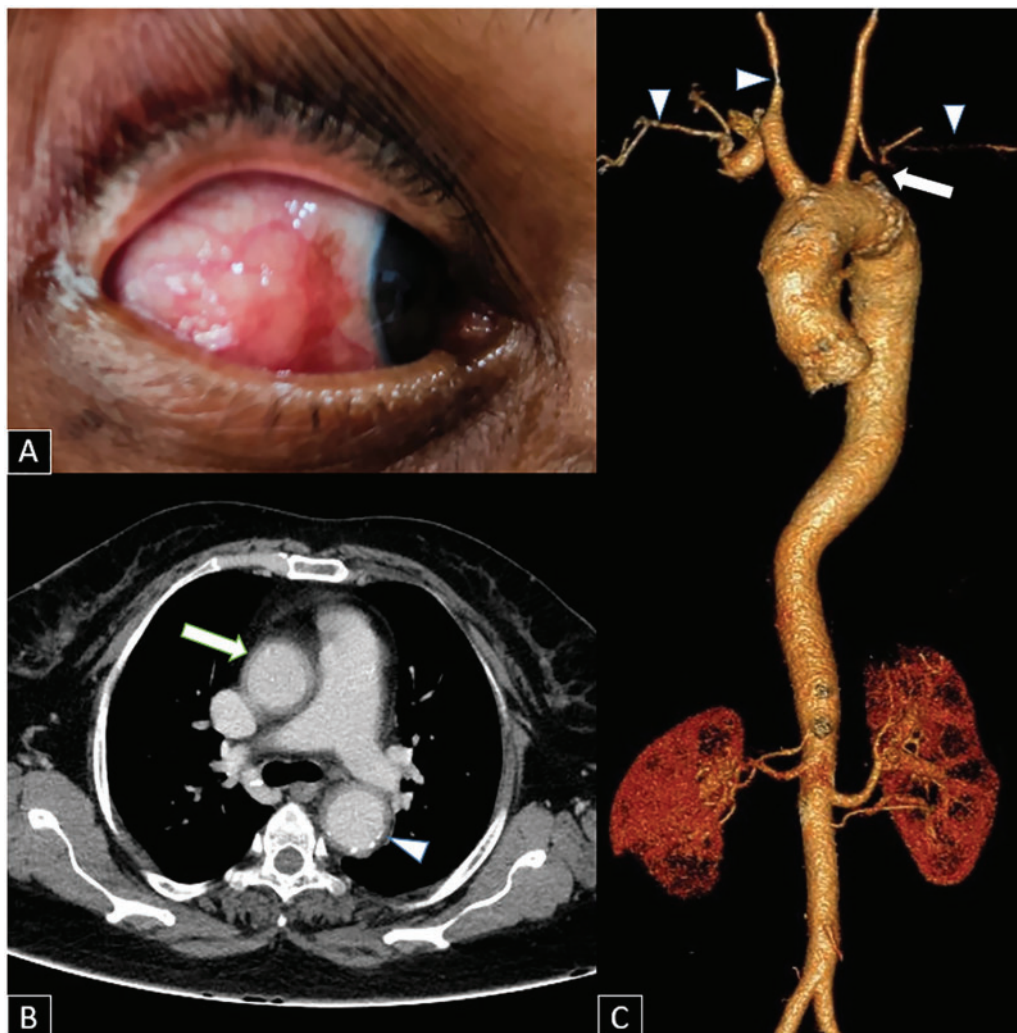
Small vessel vasculitis in Takayasu arteritis is less well described. Different cutaneous manifestations with histopathological evidence of vasculitis have been reported in Takayasu arteritis [9]. Scleritis in our patient could also be a small vessel manifestation of Takayasu arteritis. However, the possibility of these two conditions coexisting cannot be excluded.

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#### Data availability statement

Data are available upon reasonable request by any qualified researchers who engage in rigorous, independent scientific research, and will be provided following review and approval of a research proposal and Statistical Analysis Plan (SAP) and execution of a Data Sharing Agreement (DSA). All data relevant to the study are included in the article.

**Fig. 1** Scleritis in Takayasu arteritis

(A) Anterior scleritis. (B) CT angiogram, showing circumferential wall thickening in ascending (arrow) and descending thoracic aorta (arrowhead). (C) CT angiogram, showing stenosis in right common carotid and bilateral subclavian arteries (arrowheads), with complete occlusion in proximal part of left subclavian artery (arrow).

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