

Case Report

Scleritis and Takayasu's disease – rare combined presentation

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Abstract

Introduction: Takayasu's disease (TD) (pulseless disease, aortic arch syndrome) is a rare but potentially life-threatening chronic giant cell vasculitis. The major complications attributed to the disease include Takayasu's retinopathy, secondary hypertension, aortic regurgitation, and arterial aneurysm. The aim of this study is to describe a patient with Takayasu's disease and scleritis and to show its association.

Case report: We describe a 44-year-old female patient with Takayasu's disease who presented with bilateral anterior scleritis in the form of painful red eye. Patient did not have any associated autoimmune diseases. Patient was diagnosed to have TD and was treated with immunosuppressant. **Conclusion:** Scleritis though rare can be a vasculitic feature of TD. This is a first report of Takayasu's disease with scleritis, which has been documented in Nepal.

Key word: Takayasu's arteritis, scleritis, systemic immunosuppression, Computed tomography aortogram

Introduction

Takayasu's Arteritis (TA) is a rare chronic obliterative vasculitis affecting the aorta and its major branches. Takayasu's arteritis is an inflammatory arteritis involving the large arteries (Panja & Mondal 2004). Takayasu, a Japanese ophthalmologist, was the first to describe the disease in 1908. (TP 1989) It is more commonly seen in females of reproductive age and is more prevalent in Asian and Latin American countries. Takayasu's disease has an incidence of 2.6/million/year with a female to male ratio of 9:1. (TP, Bleck ,

1989) Although the pathogenesis has not been entirely elucidated, TA is considered to be a T-cell-mediated granulomatous vasculitis. It is now recognized that the inflammatory process of the large arteries affects regions of the walls supplied by the vasa vasorum, suggesting that primary small vessel involvement may contribute to the development of the clinico-pathological features of Takayasu's aorto-arteritis. Classical ophthalmic features of the disease result from reduced ocular perfusion, which manifests as hypoxic retinal changes such as microaneurysms, arteriovenous anastomosis and non-perfused areas. (Sushmita Kaushik, 2005) Smith and Rosenbaum suggested a real association between Takayasu's arteritis (TA) and scleritis because of the strong temporal

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relationship between the two conditions in their patient. (Shukla et al. 2016) Jain et al have published a report that describes a case of TA occurring in association with scleritis.(Jain et al. 2000)

Case Report

A 44 year old, female patient, nursing staff by occupation presented to our hospital with chief complaints of Redness ,pain in Both eyes for 1 week (RE> LE), mild blurring of vision in RE and headache .

Figure 1: RE Scleritis

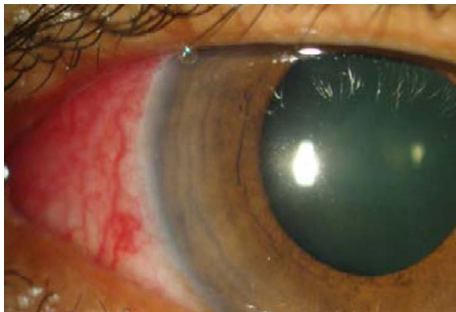
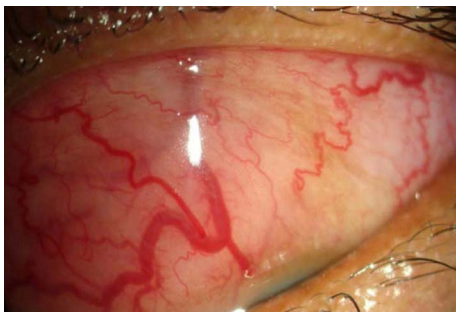


Figure 2: LE scleritis showing congestion and telangiectasia of scleral and episcleral vessels .

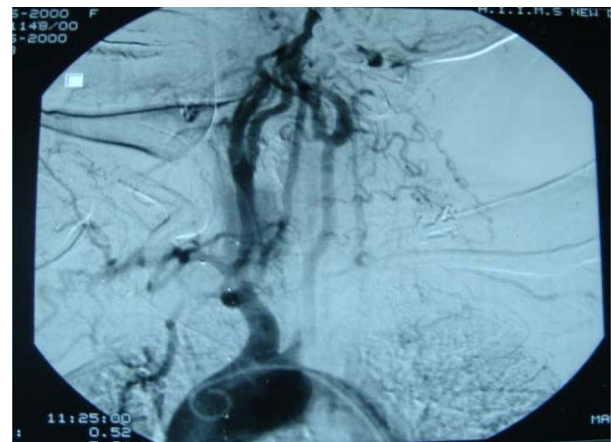


She also gives a 6-year history of fluctuating bilateral ocular redness and pain, along with intermittent numbness and stiffness in her arm and neck, palpitations, breathlessness and lightheadedness sometimes progressing to syncope, and fatigue. Medical history included anemia.

She had been investigated eighteen months earlier for both intermittent arm pain with claudication and had been diagnosed as having Bilateral Subclavian artery occlusion. She was a non-smoker and no underlying cause for the

arterial disease had been identified at that time. Her visual acuities were 6/12in RE and 6/6 for LE with mild posterior subcapsular cataract in RE and normal intraocular pressures. There was no retinal vasculitis. Fundus Examination was within normal limit. Thus, she was provisionally diagnosed as, Bilateral Non necrotizing Anterior Scleritis (as seen in Figure 1 and 2). Systemic examination revealed absent pulses in the radial, ulnar, and brachial arteries of her both arms with carotid and subclavian bruits. The right arm blood pressure (BP) measured 100/60 left arm was 120 / 70 mm of Hg and the leg BP was 180/100. An arch aortogram showed complete occlusion of the left common carotid and subclavian arteries and a tight stenosis of the proximal right subclavian artery with a mild stenosis of the right carotid artery. (As seen in figure 3)

Figure 3: Carotid arch aortogram showing Occlusion of subclavian artery



An echocardiogram showed mild mitral valve and Tricuspid valve regurgitation. Investigations following the diagnosis of scleritis included a raised erythrocyte sedimentation rate at 72 mm in the first hour with CRP - ++++ (Positive)

Antinuclear antibodies (ANA) and antineutrophilic cytoplasmic antibodies (ANCA) were negative. Other Investigations were within normal limits (Hb- 11.0 g/dl, Creatinine -0.8mg/dl, serum globulin- 6.0g/dl). With all these clinical features and investigations,

she was diagnosed as Takayasu's disease with B/L Anterior Scleritis. The patient was started on oral prednisolone 1 mg/kg bodyweight along with methotrexate 10 mg/week, with folate supplements. Prednisolone was tapered by 5 mg/week after a month till 20 mg/day and methotrexate to 7.5 mg/week, calcium supplements were also given. The patient was also given Tab. Amlodipine (10mg) daily as she was hypertensive. Her attacks of recurrent scleritis subsided (as seen in Figure 4) but right arm claudication persisted for which she is planned for right subclavian artery stenting. On follow up examination she had developed RE posterior subcapsular cataract (Figure 5) with VA of 6/36 and Diabetes mellitus. She is being treated for DM conservatively and is still waiting to undergo cataract and reconstructive arterial surgery.

Figure 4: RE and LE on follow up with regression of scleritis

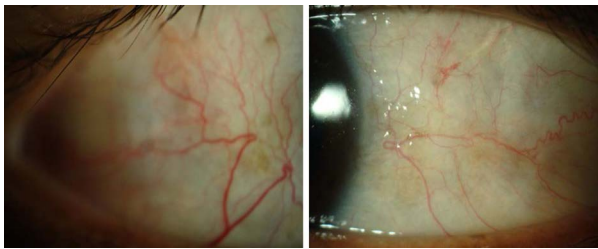
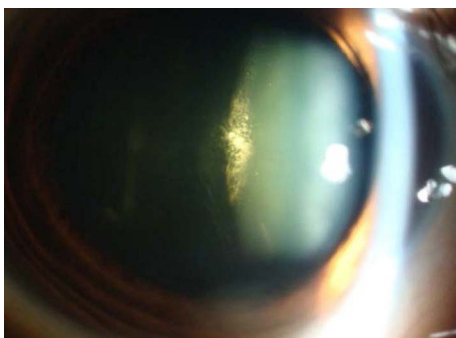


Figure 5: Posterior subcapsular cataract



Discussion

Takayasu's vasculitis generally involves large and medium sized arteries, in particular, the great vessels. A presumed T cell-mediated autoimmune inflammation of the vasa-vasorum progressing to fibrosis, and resultant arterial

stenoses, occlusions, and aneurysms causing various ischemic symptoms.(Rizzi et al1999).

Constitutional symptoms in TD include weight loss, headache, malaise, arthralgia and fever. There is carotid artery bruit in 80% cases, difference in the blood pressure of extremities, claudication, carotodynia, hypertension, congestive heart failure and aortic regurgitation. TIA, stroke, Ocular Ischemic syndrome and seizures are also associated with Takayasu's arteritis. Ocular signs of Takayasu's disease include episcleritis, corneal edema, ischemic pseudoiritis, iris atrophy, low intra-ocular pressure, neovascular glaucoma, cataract, retinal vascular changes, cotton wool spots, disc edema, exudative retinal detachment and macular edema.(Brown GC 1988)

Our patient satisfied all the 5 of the 6 American Colleges of Rheumatology (ACR) criteria for TA.(FLYNN et al. 2010). As per ACR 1990 criteria, (Arend et al. 1990) a patient is diagnosed as TA if she satisfies 3 out of 6 of the following criteria:

1. Age at disease onset <40 years
2. Claudication of extremities
3. Decreased brachial artery pulse
4. BP difference >10 mmHg between arms
5. Bruit over subclavian arteries or aorta
6. Arteriogram abnormality, arteriographic narrowing, or occlusion of the entire aorta, its primary branches or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or similar causes; changes usually focal or segmental.

Laboratory findings of anemia, raised erythrocyte sedimentation rate, and elevated serum γ globulins are prominently common. Syphilis and other autoimmune diseases clearly associated with scleritis, eg, rheumatoid arthritis and Wegener granulomatosis which are

important differential diagnosis were excluded by serological testing and other clinical and laboratory tests.

Ocular abnormalities are common in TD. The ophthalmic complications are generally late manifestations of the disease and include ischemia of the retina, choroid and anterior segment. Classical ophthalmic features of the TA result from reduced ocular perfusion, which manifests as hypoxic retinal changes such as microaneurysms, arteriovenous anastomosis and non-perfused areas. (Sushmita Kaushik, Amod Gupta, Vishali Gupta, 2005)

Characteristic Takayasu's retinal vasculopathy with potential neovascular complications occurs in a third of cases, and hypertensive retinopathy affects 20% of patients. (Kiyosawa & Baba 1998). Visual aberrations may also result from cerebral ischemia. Arteritis involving the aortic arch and its branches favors the development of ischemic ocular complications (Peter et al. 2011).

Takayasu's disease presenting as scleritis is very rare and is likely to be overlooked by the general ophthalmologist. The 5 years mortality of TA being 35%, (Rizzi et al. 1999) it is an important diagnosis which may often be apparent after thorough history and examination.

To our knowledge there is no report of association of TA and scleritis from Nepal. There are few reports from other parts of the world. Jain et al, Justine et al and Shukla et al have reported rare reports of scleritis and Takayasu's Disease. Management of TD involves systemic immunosuppression for acute lesions and surgery for fibrotic pathology. Corticosteroids reduce inflammation but are not curative. Methotrexate may be useful in inducing a remission. Our patients scleritis and constitutional symptoms responded to immunosuppressants, but surgery is required to relieve the symptoms of cicatrizing arterial stenosis. Other systemic diseases

associated with scleritis, such as Wegener's granulomatosis, have a specific serological test like ANCA to support the diagnosis. While scleritis in rheumatoid arthritis or systemic lupus erythematosus occurs in already established disease, thus is easy to treat. As opposed to that TA is suspected on clinical evaluation and confirmed on aortogram.

Hence, it is important for ophthalmologists, especially those practicing in Asian countries to be aware of the life-threatening implications of an undiagnosed and untreated episode of scleritis, especially in middle-aged Asian female population. Certain laboratory investigations such as ESR, CRP, and appropriate vascular imaging can quickly lead to an obvious diagnosis.

Although rare, we document an association between TD and scleritis. Scleritis is a feature of many of the vasculitides. This case alerts us to the possibility of Takayasu's disease presenting as a more acute inflammatory ocular picture before any signs of the chronic ischemic changes in the eye can be found.

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