Scleroderma-related choroiditis
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Abstract:
A 34-year-old female a known scleroderma patient presented to us with ocular manifestations in the form choroiditis along with optic nerve involvement. Blood investigations were done, and the ocular findings were confirmed by fundus fluorescein angiography. She was successfully managed with systemic steroid therapy. Among the protean ocular manifestations in scleroderma, only a few cases of choroidal involvement have been reported. Further, clinical and histopathological studies may be needed to throw light on the exact etiopathogenesis.

Keywords:
Choroiditis, optic nerve, scleroderma, steroids

Introduction
Scleroderma is an autoimmune connective tissue disease of unknown etiology characterized by immune activation, vasculopathy, and excess fibrosis. Ocular involvement of scleroderma is widely recognized, though choroiditis is very uncommon. We report a case of scleroderma-related choroiditis with optic nerve involvement, which responded dramatically to oral corticosteroid therapy.

Case Report
A 34-year-old south Indian female came to our outpatient department with complaints of insidious onset blurring of vision with gradual progression in the right eye for 2 months. The fall in vision was not associated with any pain or redness. She gave us a history of a similar episode of blurring in the same eye 3 years back, for which she was treated with intravenous methyl prednisolone followed by oral steroids following which her vision returned to normal.

She was diagnosed to have scleroderma a year back, following a skin biopsy from her forearm which showed features suggestive of scleroderma. At that time, she had hyperpigmentation over her ear lobules and nose and also suffered from gastritis and dysphagia. She was treated with pulse cyclophosphamide and then later shifted to oral mycophenolate mofetil (500 mg) twice a day by her rheumatologist. She was gradually withdrawn from systemic medications following the recovery of her systemic condition. She was not on any medications for more than 6 months when the current ocular symptoms started.

On ophthalmic examination, the best-corrected vision was 20/30 in the right eye and 20/20 in the left eye. Ocular motility was normal. Slit-lamp examination of the right eye showed quiet anterior chamber and anterior hyaloid face. Examination revealed normal sized pupil reacting well to light, and there was no relative afferent pupillary defect. The rest of the anterior segment examination was within normal limits. Fundus examination of the right eye showed a clear vitreous cavity with an edematous hyperemic optic disc along with multiple hypopigmented patches scattered throughout the posterior pole, suggestive of choroiditis [Figure 1]. The left eye anterior and posterior segment examination was normal.

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Her routine blood investigations showed a hemoglobin level of 10.6 g%, raised erythrocyte sedimentation rate of 38 mm/1 h. Peripheral blood examination showed hypochromic, microcytic red blood cells with occasional teardrop cells. Renal function tests which included blood urea and serum creatinine was done and the values were within normal limits. Test for antinuclear antibody was positive. Magnetic resonance imaging of the brain and orbit revealed no abnormality and a normal posterior coat of eyeball. She was also examined by our in house physician and rheumatologist, and all her systemic examination was within normal limits. Fundus fluorescein angiography (FFA) showed hypofluorescent areas in the early phase which corresponded to the hypopigmented lesions seen clinically, which increased in intensity in the late phase along with disc staining [Figure 2].

We reached on a working diagnosis of right eye choroiditis with optic nerve involvement and started her on a course of oral steroids (1 mg/kg bodyweight) to which she responded dramatically. The steroids were gradually tapered over the next 3 months during which she did not have a relapse. This time we did not initiate her on any immunosuppressive drugs.

At her follow-up at 5 weeks, the right eye fundus lesions resolved and the vision improved to 20/20 [Figure 3]. The patient is on a regular follow-up with us and rheumatologist for the last 2 years, with a stable vision of 20/20 in both her eyes, with no episodes of recurrence. Her systemic condition is also stable and is currently sans all medications.

**Discussion**

Scleroderma is a chronic multi-system disorder of unknown etiology characterized by immune activation, vasculopathy, and excess fibrosis.\(^1\) Activation of the immune system (cellular and humoral) appears to be pivotal to the pathogenesis of scleroderma. Ocular involvement of scleroderma ranges from the anterior to posterior segment, the orbit, and the extraocular muscles. Eyelid skin abnormalities and keratoconjunctivitis sicca are the most common scleroderma-related ocular findings.\(^2\) In the posterior segment, the most commonly reported ophthalmoscopic findings include microvascular changes, cotton-wool spots, retinal exudates, retinal and optic nerve head edema, retinal hemorrhages, retinal vein, and artery occlusion.\(^3,4\) However, there is a paucity of reports on the choroidal changes in scleroderma.

FFA studies in patients with scleroderma have demonstrated abnormalities of the choroidal circulation.
and retinal pigment epithelium.\textsuperscript{[5‑7]} Grennan and Forrester,\textsuperscript{[6]} in an FFA study of 10 patients with scleroderma, found 50% of the patient to have choroidal hypoperfusion affecting the choriocapillaris and small arterioles.

Histological studies\textsuperscript{[6,10]} in patients with scleroderma have shown that the choroidal vessels are grossly affected with endothelial cell damage, basement membrane thickening, and absence of pericytes and deposition of abnormal material in and around the endothelium. Similar structural alterations are known to be present in affected skin, normal-appearing skin of patients with localized scleroderma, and in the renal vessels in those patients suffering from generalized scleroderma. The retinal and choroidal vasculatures appear to provide an almost ideal window through which to study the generalized arteriolar and capillary pathology of scleroderma.

Our patient with scleroderma developed choroiditis with secondary optic nerve involvement. Clinical testing and investigation could not identify an underlying etiology other than scleroderma. Being an uncommon manifestation and due to the paucity of histopathological evidence, the exact pathogenesis and etiology still remains unanswered. The choroidal changes can be presumed to be due to vascular damage of the choriocapillaris. The unilateral involvement in our case can be because of the localized pathological changes as seen in localized scleroderma, which are more common in childhood and young adults, and are almost never associated with significant systemic involvement, in contrast to a systemic variant of this clinical entity. The absence of inflammation in vitreous can be explained partly with noninflammatory proliferative or obliterative nature of vasculopathy seen in cases with long-standing scleroderma.\textsuperscript{[11]} Choroidal involvement is an uncommon occurrence in scleroderma, with only a few cases reported. The mechanism for this manifestation remains known, but the use of systemic steroids in our patient resulted in control of the process suggesting that an inflammatory mechanism plays a significant role. However, further clinical studies and histopathological confirmation may be needed to support this.

\textbf{Declaration of patient consent}

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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\textbf{Conflicts of interest}

There are no conflicts of interest.

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