

## Unilateral acquired Brown's syndrome in systemic scleroderma: An unusual cause for diplopia

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Brown's syndrome can be congenital or acquired with multiple causes. It has been described as an ocular complication in various rheumatic and nonrheumatic diseases. We describe a case of 27-year-old female patient with 5 years old history of systemic scleroderma who developed vertical diplopia, a left head tilt, and restriction of left eye on elevation in adduction. The patient responded to systemic steroids with resolution of diplopia.

**Key words:** Brown's syndrome, corticosteroids, systemic scleroderma, vertical diplopia

Brown's syndrome is an ocular motility disorder characterized by the inability to fully elevate the affected eye in adduction. The acquired variety involves secondary changes in a previously normal superior oblique tendon or tendon trochlear complex. Acquired Brown's syndrome cases have been reported in rheumatological and nonrheumatological disorders.

### Case Report

A 27-year-old woman, presented with sudden onset of diplopia since 1 day, which was greatest in the right side. She had 5 years old history of systemic scleroderma (systemic sclerosis) with Raynaud's phenomenon.

She was previously treated with oral prednisolone and was not on any steroid treatment when she presented to us with the complaint of diplopia. She had numerous extraocular

findings mainly cutaneous (flexion contracture, limited joint mobility, calcinosis, Raynaud's phenomenon, sclerodactyly, telangiectasias, microstomia) [Fig. 1a and b]. She had no other systemic manifestations.

Her uncorrected visual acuity was 20/20 in the right eye (RE) and 20/30 in the left eye (LE). Best corrected visual acuity was 20/20 in both eyes (LE - 0.5 DC × 180°). Pupillary responses and other ocular examination were normal. There was a slight exophoria in the primary position. Diplopia was present on upgaze, most pronounced in right gaze, with minimal diplopia on right downgaze. Elevation of the LE was correspondingly limited in adduction, with minimal limitation of depression in adduction [Fig. 2a]. There was no limitation of elevation in abduction.

Hess chart was consistent with Brown's syndrome [Fig. 3a]. This appearance of the Hess chart taken with the clinical picture is diagnostic of restriction of the superior oblique tendon (Brown's syndrome). All other movements were intact.

Exaggerated forced duction test was done under local anesthesia which confirmed the mechanical restriction. Fundus examination was normal in RE and showed intorsion in LE



**Figure 1:** (a) Skin thickening of the fingers extending proximal to the metacarpophalangeal joints and severe flexion contractures of the fingers. (b) Flexion contractures of toes

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[Fig. 4a and b]. There was no evidence of torsion by subjective test. Magnetic resonance imaging (MRI) brain and orbits were normal.

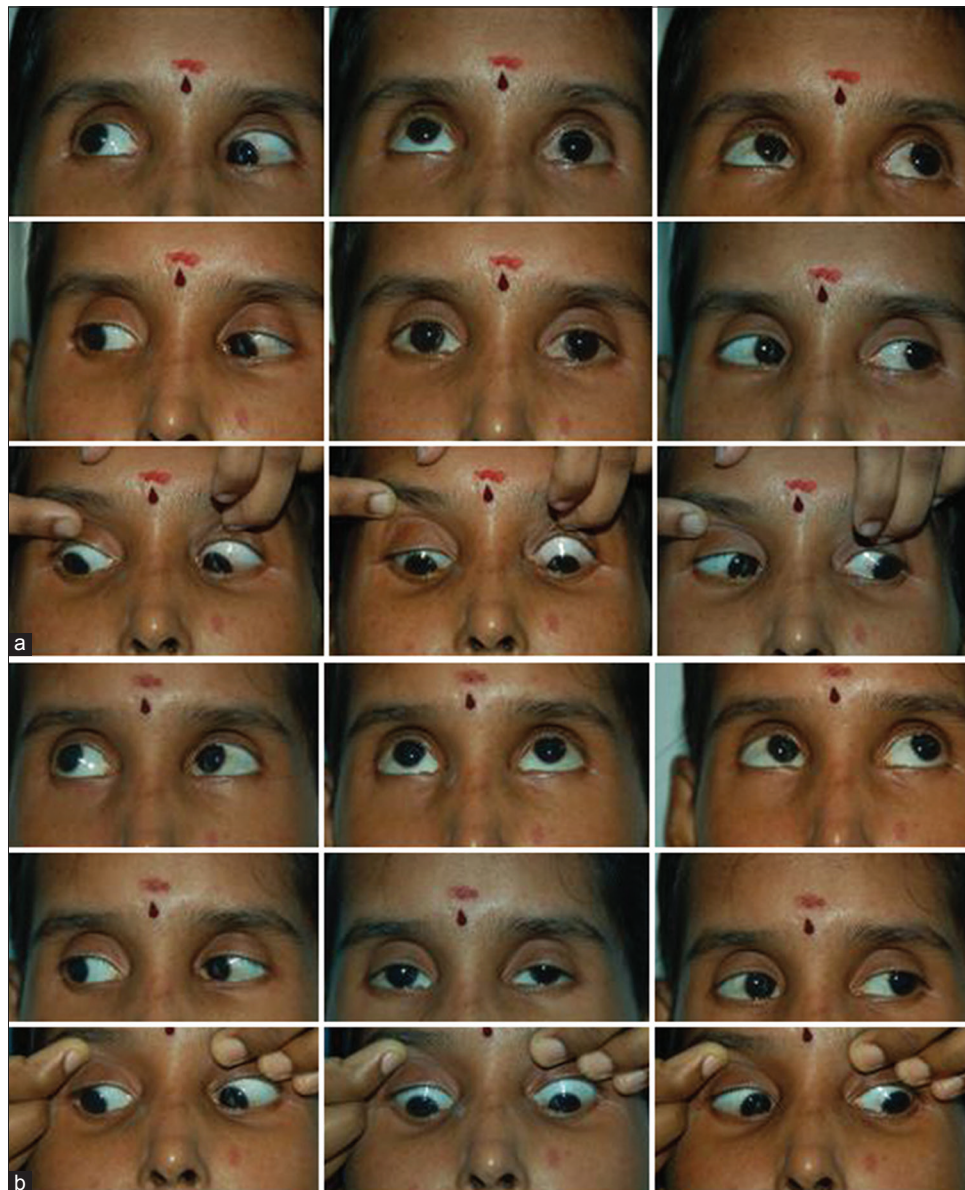
Laboratory studies showed a hemoglobin concentration of 12.5 g/dL, a leukocyte count of 13,500 cells/ $\mu$ l, and erythrocyte sedimentation rate of 20 mm/h (Westergren). Total blood count and glucose concentrations were normal.

Antinuclear antigen blot profile revealed antibodies to double stranded DNA and antitopoisomerase-1 anti-bodies (anti-Scl-70) to be positive. This patient met the criteria developed by the American College of Rheumatology and the European League Against Rheumatism leading to definitive diagnosis of systemic scleroderma.<sup>[1]</sup> She was given prednisolone 20 mg daily for 1 week which was tapered to 10 mg and 5 mg subsequently for 1 week each after physician opinion. She came after 10 days with resolution of the diplopia in primary gaze and diplopia in upgaze only. On follow-up

after 1 month, she had no diplopia with complete resolution of limitation of elevation on adduction,[Fig. 2b] which was confirmed by Hess charting [Fig. 3b]. After 4 months of follow-up, she was stable with no diplopia in any gaze.

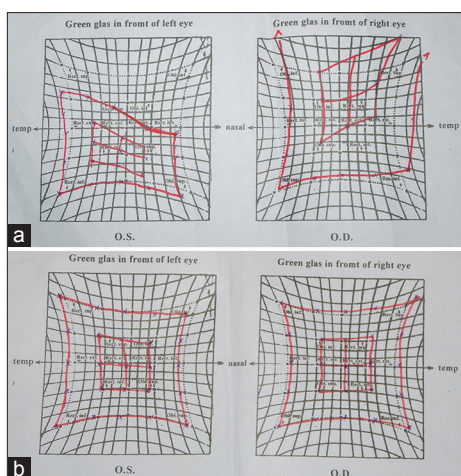
## Discussion

Systemic scleroderma (systemic sclerosis) is an autoimmune or connective tissue disease. It is characterized by thickening of the skin caused by accumulation of collagen and by injuries to the smallest arteries. Skin thickening of the fingers extending proximal to the metacarpophalangeal joints is sufficient for a patient to be classified as having scleroderma.<sup>[1]</sup> Acquired Brown's syndrome cases have been reported in rheumatological and nonrheumatological disorders.<sup>[2-6]</sup> Tenosynovitis in rheumatic disorders may affect ocular muscles producing symptoms of diplopia. If the superior oblique tendon cannot lengthen or slide freely, the affected eye



**Figure 2:** (a) Versions in the nine cardinal gazes at the time of presentation. There is under elevation in adduction of left eye which is worse in upper right gaze. (b) Versions in the nine cardinal gazes after 1 month. There is no limitation in upgaze in adduction of left eye





**Figure 3:** (a) A Hess chart at presentation suggestive of Brown's syndrome. (b) Hess chart 1 month later showing pronounced improvement with minimal underaction of superior oblique in left eye

cannot be elevated completely in full adduction. The patient reports diplopia on upward gaze. The Brown's syndrome has been also described in association with various nonrheumatic disorders including frontal sinusitis or frontal sinus surgery, blepharoplasty, trauma, focal metastatic lesions, preseptal cellulitis, hypogammaglobulinaemia, Hurler Schies, combined lichen sclerosus et atrophicus and morphea, psoriasis.<sup>[2-9]</sup>

It has been suggested that the pathophysiology of rheumatic diseases associated to Brown's syndrome is a superior oblique impingement secondary to stenosing tenosynovitis, similar to trigger finger.<sup>[4]</sup> The diagnosis is mainly clinical and imaging tests such as MRI may show inflammation of the superior oblique tendon or trochlea. However, this finding is not always present.<sup>[3,4]</sup>

In our patient, local tenosynovitis may be the underlying mechanism responsible for Brown's syndrome. The superior oblique was normal in MRI. The resolution of symptoms within a month associated with the systemic use of steroids; however, points to a tenosynovitis of the superior oblique tendon as the cause of diplopia in our patient.

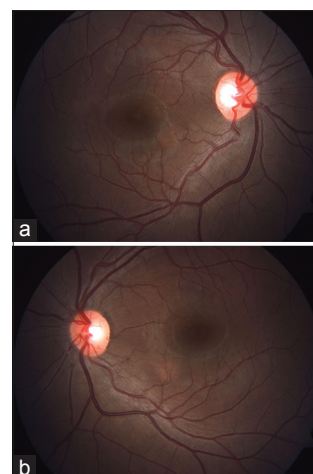
The literature contains only one case report about bilateral Brown's syndrome in systemic scleroderma which was treated by local administration of steroids.<sup>[10]</sup>

Our case is different from the case reported by Gezer *et al.* as their case was bilateral Brown's syndrome in a 56-year-old female who responded to prednisolone injection 1 mL (40 mg) into the superonasal aspect of the orbits whereas our case was unilateral Brown's in young female who responded to systemic steroids.

This case report illustrates systemic scleroderma (systemic sclerosis) as another potential cause of acquired Brown's syndrome and role of ophthalmologist in its management.

#### 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,



**Figure 4:** (a) Fundus picture of right eye. (b) Fundus picture of left eye showing intorsion

and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

#### Conflicts of interest

There are no conflicts of interest.

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