Photo Essay

Localized scleroderma causing enophthalmos: A rare entity

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Key words: Enophthalmos, orbit, scleroderma

A 27-year-old lady presented with painless progressive shrinking of left eye since 20 years [Fig. 1a and b], following orbital trauma with iron rod. She also had a left inguinal skin lesion [Fig. 1c] since last 15 years. There was no history of any orbital surgery or any systemic symptoms. On ocular examination, her visual acuity was 20/20 both eyes. Slit lamp examination both eyes were normal except for few corneal glassy discrete endothelial precipitates [Fig. 1d]. Hertel's exophthalmometry measured 15 mm and 10 mm in right and left eye, respectively. Periocular sensations were normal on both sides. She had a band like diagonal line on left forehead that is a form of localized scleroderma also known as "en coup de sabre" [Fig. 1a - black arrow] because of its resemblance with a sword wound. However, both sides of the face looked symmetrical. Fundus examination in left eye revealed retinal folds in macula [Fig. 1e]. Axial length in both eyes was 21 mm. Computed tomography scan orbit showed gross enophthalmos in the left eye with reduced retrobulbar fat volume with no sign of any orbital fracture or any orbital mass [Fig. 1f and g]. On general examination, there was no palpable breast lump, but she had sclerotic skin lesions in left inguinal region that had been diagnosed as morphea type of localized scleroderma on skin biopsy by a dermatologist depending on presence of sclerosis of the skin and underlying tissues owing to excessive collagen deposition, adnexal atrophy, and mononuclear cell infiltrates. Systemic work up and autoantibody profile were negative. Intraorbital hyaluronic acid gel injection or autogenous fat transfer was suggested for orbital volume augmentation. However, she preferred observation and was not keen for any intervention. At 3 years follow-up, patient was status quo. Localized scleroderma, which manifested in both forms of linear scleroderma and morphea in this case, involved the orbit causing orbital fat atrophy leading to enophthalmos. Absence of hemifacial atrophy ruled out the diagnosis of Parry-Romberg syndrome in this case.

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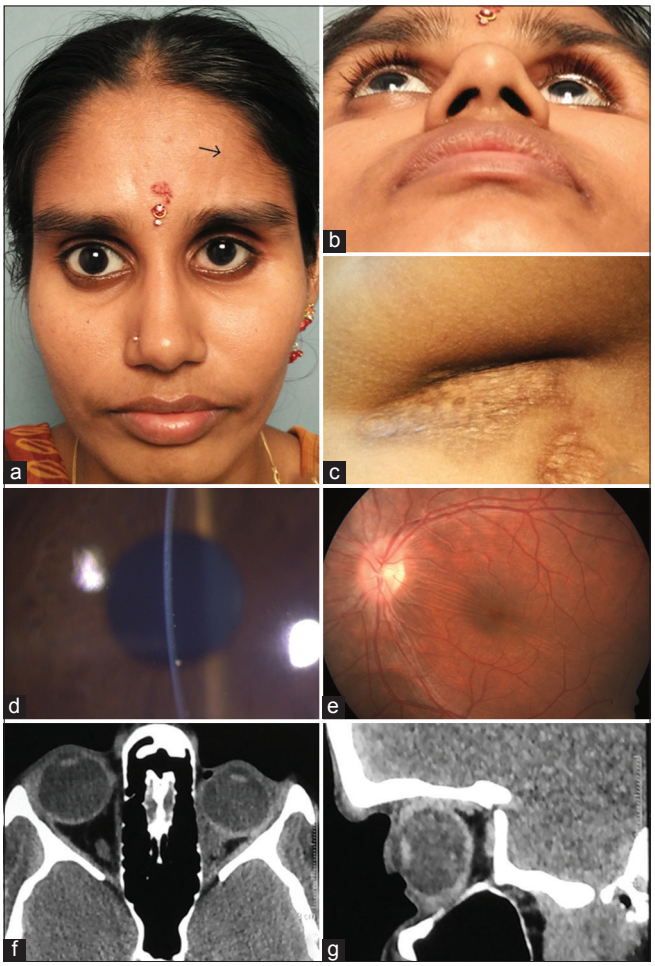

Figure 1: (a and b) Clinical photographs showing gross enophthalmos of left eye with linear diagonal scar over left forehead (Coup de Sabre-black arrow); (c) atrophic and sclerotic skin lesion in left inguinal region; (d) slit lamp photograph of left eye showing corneal endothelial precipitate; (e) fundus photograph of left eye showing multiple retinal folds at macula; (f and g) computed tomography scan orbit (axial and saggital section) showing enophthalmic left eye with reduced retrobulbar fat volume with absence of any other abnormality

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Discussion

Scleroderma refers to a heterogeneous group of autoimmune fibrosing disorders. The disease can be either "localized" to the skin or "systemic" involving other organs in addition to the skin. Localized scleroderma can be further divided into linear and morphea (localized or generalized patches of atrophic and sclerotic skin lesions).[1] Linear scleroderma is characterized by localized fibrosis of skin, blood vessels, subcutaneous fat, muscle, and sometimes bone. It primarily affects the pediatric population in the first and second decade.[2] When linear scleroderma occurs on the fronto-parietal scalp, it is referred to as "'en coup de sabre," given the resemblance of the skin lesion to the stroke of a sabre.[3] Specific neurological and ophthalmic associations of craniofacial scleroderma have been reported in literature that were all ruled out in our patient. The ophthalmic manifestations in scleroderma may include ptosis, lid induration or tightening, lash loss, motility disorder, keratoconjunctivitis sicca, conjunctival injection and vascular sludging, fornix shortening, iritis, heterochromia iridis, papillitis, retrobulbar pain, enophthalmos, displacement of outer canthus from resorption of orbital bone, and retinal hemorrhages.[2-5]

Conclusion

Scleroderma is a rare cause of enophthalmos after ruling out other likely differentials of orbital fracture and orbital metastatic breast cancer. The linear scar on the forehead "en coup de sabre" should not be discarded as an innocuous finding rather should aid the examiner in making the correct diagnosis.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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