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Linear scleroderma 'en coup de sabre' with ptosis and oculomotility disorders: case report and review of the literature

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Abstract

Aim: **To present a case of linear scleroderma (LS) involving the extraocular muscles and periorbita with follow-up over an 18-year period.**
Method: **Initial presentation was of a female aged 9 years with progressive left unilateral ptosis which ultimately underwent surgical correction.**
Results: **The main features of 'en coup de sabre' subsequently manifested on the left side of the face and head. An unusual feature was the characteristic subcutaneous cleft which also appeared on the opposite side above the right eyebrow. The left eye went on to develop a corneal ulcer with resulting poor visual acuity. An ipsilateral oculomotility disorder was then noted in addition to poor eyelid closure.**
Conclusion: **This case highlights the need to exclude 'en coup de sabre' as a potential differential diagnosis in any case of progressive or recurrent ptosis, especially in childhood. As oculomotility may also be affected ultimately, it could lead to corneal compromise following ptosis correction.**

Key words: Key words: 'En coup de sabre', Oculomotility disorder, Ptosis, Scleroderma

Introduction

Linear scleroderma (LS) is an uncommon autoimmune disease which presents clinically as a 'localised' form in which purely local areas of skin are affected or a 'systemic' form where internal organs in addition to the skin are affected. Lesions usually start with contraction and firmness of the skin over the affected area. An ivory irregular sclerotic plaque can subsequently develop, sometimes with telangiectatic vessels coursing over it together with hyperpigmentation at the edge.[1] Scleroderma 'en coup de sabre' is a term used to describe a lesion of LS which is specifically localised to the frontal parietal section of the face and scalp. A linear depressed groove appears on the frontoparietal region extending into the scalp producing a linear zone of alopecia, which may be preceded by a bleaching of the hair. Not infrequently there is atrophy of the corresponding part of the face and cheek, with facial asymmetry which usually occurs within a year.[1] This condition may also be associated with atrophy of one side of the face. 'En coup de sabre' is an uncommon and seldom reported dermatological disorder; it is usually unilateral, but rare bilateral cases have been recorded.[7] It is often reported with ptosis or lid abnormalities[4–9,11–16] but concurrent oculomotor disorders are rare.[4,7,9,11,13–16]

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Case report

In 1991 a female patient aged 9 years presented to the Ophthalmology Department. Her unaided visual acuities were 6/5 in each eye using a standard 6 m Snellen chart and orthoptic assessment proved her to be fully binocular. She had a progressive left ptosis, which had developed over a period of 6 months. This was associated with left periorbital swelling and some discoloration of the skin around the eye. Her ptosis measured 2 mm with 13 mm of levator function bilaterally.

Her left pupil was noted to be slightly smaller than the right but this was thought to be a simple anisocoria. Photographs of the patient aged 1 to 9 years were reviewed and this indicated that the ptosis had been present since approximately 4 years of age. A full orthoptic examination was undertaken which showed no additional abnormality.

A referral was made to the paediatric neurologists. Subsequent assessments and investigations, including CT scan of the brain and orbits and a chest X-ray, were unhelpful in discerning the aetiology of the problem.

The degree of ptosis increased over the next 3 months to measure 8 mm with 8 mm of levator function. It remained 16 mm on the right. A decision was made to proceed with ptosis surgery and the patient underwent an anterior approach left levator palpebrae superioris resection of 16 mm. Post-operatively 8 mm of ptosis remained.

An exploration of the eyelid was carried out 5 months later and reattachment of the aponeurosis performed. Post-operatively the ptosis reduced slightly to 5 mm.

The patient was referred for a second opinion to a national tertiary referral oculoplastics service. A diagnosis of 'late acquired levator muscle dystrophy' was made. Suggested management included either a repeat levator muscle resection (with or without a muscle

Fig. 1. The patient aged 12 years showing the linear band of alopecia along the left side of the scalp. (Photograph obtained with the patient's and parents' informed formal consent.)

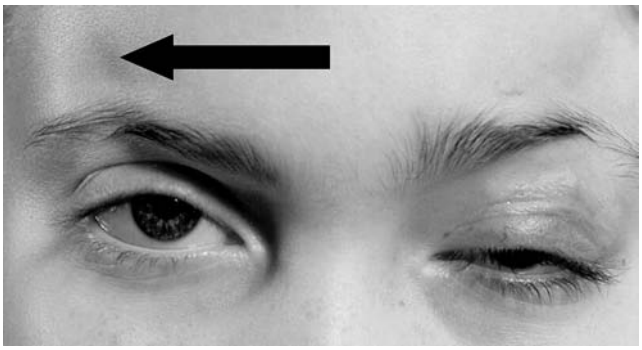

Fig. 2. The patient aged 12 years, after three surgical corrections of her left ptosis. Note the subcutaneous cleft that has appeared on the opposite side above the right eyebrow. (Photograph obtained with the patient's and parents' informed formal consent.)

biopsy) or a brow suspension. During this time, though, the patient began to complain of a linear band of alopecia along the left side of the scalp (Fig. 1) and her mother had noticed a cleft in her forehead (Fig. 2). The opinion of a dermatologist was sought and the patient was diagnosed with linear morphea ('en coup de sabre'). She commenced a course of ciclosporin which had no effect on her ptosis or her morphea.

A left brow suspension was performed 2 years later which initially improved cosmesis for 12 months but then deteriorated again to leave 7.5 mm of ptosis. The brow suspension was repeated at the age of 15 years, again with a good cosmetic result, and the patient was discharged from regular follow-up.

The patient presented as an emergency aged 21 years with a left corneal ulcer and reduced corneal sensation. Unaided visual acuities were 6/5(right) and 6/36(left). Several treatments were attempted for the left corneal ulcer, including antibiotic and antiviral medications and a bandage contact lens. These were unsuccessful and a tarsorraphy was suggested. The patient refused further surgery and then failed to attend several appointments and was subsequently discharged.

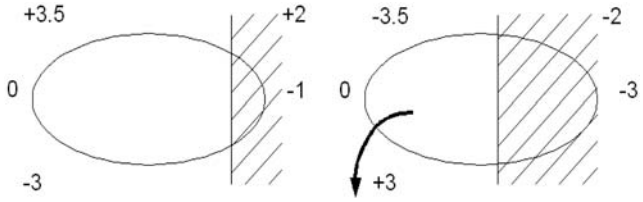

Fig. 3. Ocular motility examination included a moderate limitation of the left eye on abduction and slight limitation of the right eye on adduction.

A new referral was received 4 years later. The patient, now aged 25, was aware of a significant deterioration in her left vision. Visual acuity with a slight myopic correction was Right–0.1 (6/4.8 Snellen equivalent) and Left 0.7 (6/30 Snellen equivalent) using logMAR. A central corneal scar was noted on her left eye, with inferotemporal corneal vascularisation. There was a central band of punctate epithelial erosions. Reduced corneal sensitivity of the left eye was confirmed. There was 2–3 mm of lagophthalmos with a central eyelid notch, lacking in eyelashes. The lid was difficult to evert. There was no levator function. On the right, levator function was normal. An abnormal head posture was detected and a referral made to the orthoptic department. Examination revealed significant duction deficits of ocular motility (Fig. 3) including moderate limitation of the left eye on abduction and slight limitation of the right eye on adduction. A moderate left hypotropia with a small left divergent strabismus without diplopia (due to left suppression) was present. A CT scan of the head with contrast showed the left medial and inferior rectus muscles to be much thicker than on the contralateral side and the rest of the ipsilateral recti muscles. This suggests that restriction may account for the duction deficits observed (Figs. 4 and 5). There was also a subtle linear enhancement in the left posterolateral inferior cerebellum, consistent with a deep venous anomaly. An MRI scan of the brain and orbits carried out 9 months later revealed that the left medial and inferior rectus had decreased in size and now had a 'slightly thinner than normal' appearance (Figs. 6 and 7), but the ocular motility remained unaltered.

The patient declined further surgical intervention and decided to continue on ocular lubricants alone to the left eye. She failed to attend for further follow-up.

Discussion

LS does not follow a set pattern of progression and can be associated with many different ocular abnormalities. Several published articles have looked at the ocular impact of this disease[2–16] but no reviewed papers have documented the natural progression of LS and 'en coup de sabre' for as long as in this case.

LS is believed to predominantly affect the paediatric population,[2] with 67% of patients diagnosed before 18 years of age.[3] In 2007 a large study of 750 patients with localised juvenile scleroderma was reported.[4] It identified 24 patients with significant ocular involvement, including eyelid and eyelash abnormalities, ectropion,

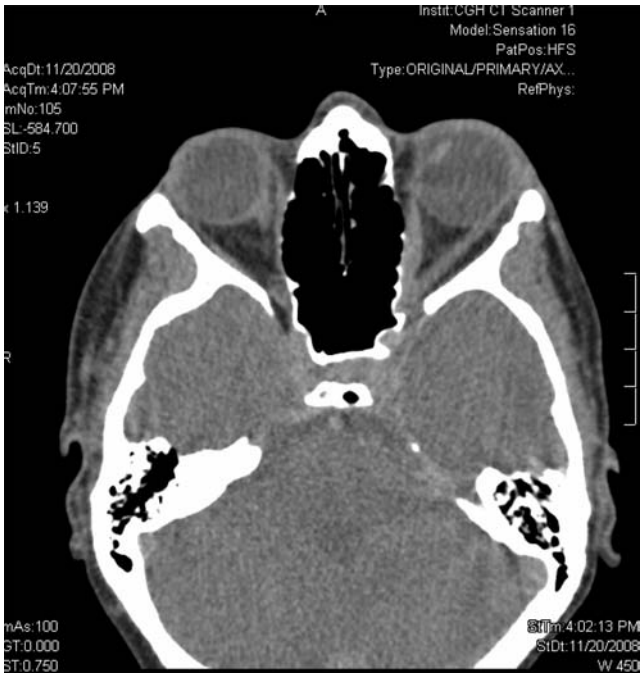

Fig. 4. Axial CT scan (post-contrast, soft tissue windows) showing thickening of the left medial rectus.

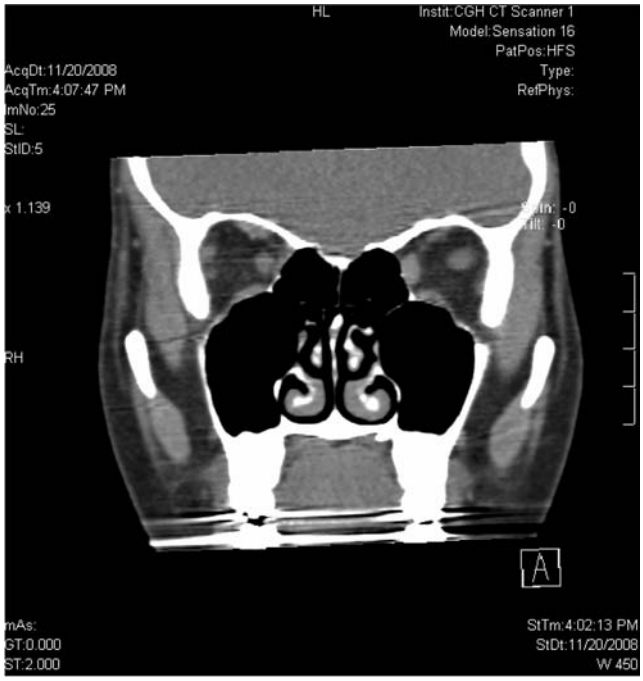

Fig. 5. Coronal CT scan (post-contrast, soft tissue windows) showing thickening of the left medial and inferior recti.

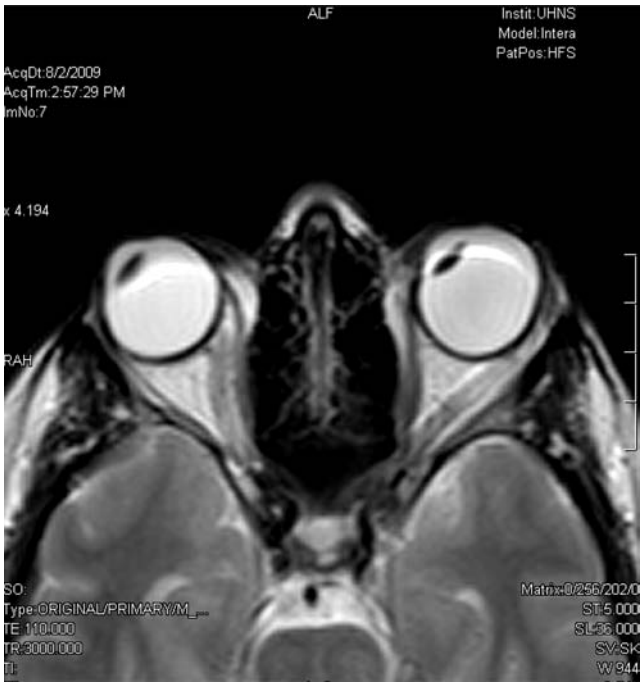

Fig. 6. Axial T2-weighted MRI scan of the orbits showing some thickening of the left medial rectus but less than on the previous CT scan.

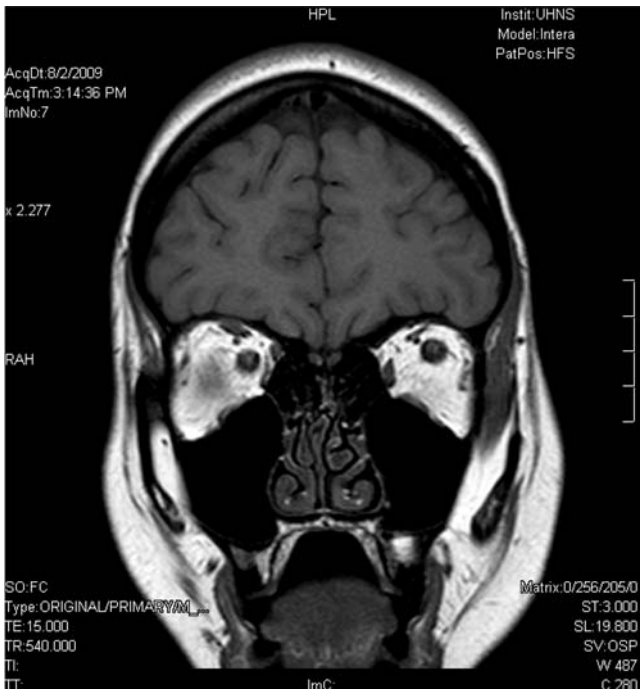

Fig. 7. Coronal T1-weighted MRI scan of the orbits showing mild thickening of the left medial and inferior recti but less than on the previous CT scan.

anterior uveitis, episcleritis, refractive error, pupil mydriasis and enophthalmos. One patient had a paralytic strabismus due to a 6th nerve palsy. Sixteen of these patients were reported to have 'en coup de sabre'.

A comprehensive literature review suggests that the lower the age of onset of LS, the greater the ophthalmic involvement. Several reports of older patients with scleroderma[5,6] identified ocular anomalies, including lid changes, conjunctival changes and lacrimal disturbances. Many of these changes were disregarded, being attributed to the natural ageing process of the eye and ocular structures rather than to scleroderma. No patient was found to have extraocular muscle involvement.

Examining this case and the literature, both the onset of bilateral LS[7] and ocular muscle involvement seems to start in the 10- to 20-year age range.[11,13,15] Females appear to develop LS at least 3–8 times more frequently than males.[17]

Areas involved in LS 'en coup de sabre' do not usually cross the midline of the body. However a case report in 2001[7] documented a 23-year-old woman with bilateral LS 'en coup de sabre' and facial atrophy. She had a complete ptosis of the left eye, a demonstrable cleft above the right eyebrow and rarefication of the left eyebrow. Neurological examination showed a facial nerve palsy and a complete oculomotor nerve palsy. In our reported case also the midline was crossed.

Neurological abnormalities were the presenting feature of a few paediatric patients with 'en coup de sabre'[3,8–10] and their cases illustrate the variability in the neurological manifestation of this condition. In one such case[8] a child presented with a dilated pupil and mild ptosis. It is unusual to have pupil dilation from an oculomotor paresis without a disturbance of the ocular motility of the other eye, and therefore other diagnoses were sought. A diagnosis of LS was eventually made.

Ptosis is a common presenting feature in 'en coup de sabre'[4–9,11–16] but ocular motility disturbances have been described in only a very few articles on LS.[4,7,9,11,13–16] These have been due to either direct muscle infiltration or peripheral nerve disease. In scleroderma, increased collagen deposition occurs, resulting in dermal thickening. Chung *et al*.[10] speculated that this disorder may progress to include other ocular findings as the dense collagen deposition within the dermis may extend to deeper connective tissue including muscle and even bone.

The aetiology of LS is unknown. It is not a hereditary disorder. David *et al.*[9] proposed that possible causative factors can include trauma, infection or severe psychological stress. Serup *et al.*[11] described a case involving the right side of the face associated with myopathy of the ipsilateral eye muscles (levator palpebrae and superior rectus). They hypothesised that a predisposition to the disease might be laid down in the mesenchyme in early foetal life before the differentiation of the anatomical structures.

There does not appear to be a causative factor in our case. The smaller left pupil was felt to be a simple anisocoria rather than a Horner's syndrome. It has been suggested that neurogenic features associated with LS could result from hyper- or hypoactivity of the sympathetic nervous system.[7]

Gambichler *et al*.[7] suggested localised forms of scleroderma may be related, belonging in the collagen-vascular group of diseases with autoimmune phenomena; or LS may be simply a developmental disease. Other theories advocate that viral or bacterial infections and genetic factors may play a role in the aetiology.[7]

In a case similar to ours, a boy presented at age 4 years with a swollen and painful, red left eye.[9] He went on to develop a groove extending from his crown to his orbital margin. A CT scan revealed mild left proptosis and a biopsy showed prominent lymphocytic infiltrate between muscle fibres in the lateral rectus. The histological findings of LS differ from those of hemiatrophy, as with sclerosis of connective tissue as a primary phenomenon in facial hemiatrophy. The marked intraocular inflammation, mild proptosis and infiltration of the rectus muscle in the patient of David *et al*. [9] supported the notion of extensive inflammation in LS; in contrast in morphea only skin is seen to be affected.

Obermoser *et al.*[12] described the first known case of 'en coup de sabre' complicated by orbital involvement to be successfully treated with interferon-γ. Intracerebral and orbital lesions had developed after two decades of the disease in this patient. The 40-year-old woman presented with 'en coup de sabre' on the left aspect of her forehead; there was also a smaller depressed skin lesion with scarring alopecia on the left parietal scalp and a smaller second band-like lesion on the right occipital scalp. Her vision was initially reported as equal in both eyes but the left vision subsequently deteriorated. An MRI scan identified a mixed cystic gliotic lesion. The vision in the left eye continued to deteriorate with associated retrobulbar pain. The lateral aspect of the left eye appeared distorted and compressed by a fibrotic stricture extending down from the forehead lesion. Examination showed a pale, atrophic optic nerve papilla and intraocular fatty tissue was also markedly reduced in the left eye, unlike in our patient. No ocular motility abnormalities were described.

There have been other attempts to treat LS but most prove to be ineffective or it is unclear for how long the treatment is effective.[9,13] One patient has been treated successfully with steroids and penicillamine; however, due to the very short follow-up it is not clear whether this improvement was permanent.[9] In another case of ineffective treatment cortisone therapy and radiotherapy was initiated but the patient complained of increasing pain around the right eye and became 'blind' on the right side.[13] Our patient did not have any response to treatment with ciclosporin. However, this intervention was commenced 2½ years after onset of disease and 1 year after diagnosis.

There are three cases reported in the literature with actual documented extraocular muscle involvement. One case documents a 21-year-old woman with LS who presented with progressive narrowing of the right palpebral fissure.[13] Her CT findings showed right superior rectus, right superior oblique and right medial rectus enlargement, with biopsy demonstrating muscular and fibroid tissue without signs of inflammation. Visual fields were normal, but colour vision was defective in the right eye. This case supports the idea that a natural stage of this disease is muscle enlargement which may be demonstrable on CT testing.

A second case report details a 51-year-old man with systemic scleroderma who had tight, thickened skin of the face and lids which gave the appearance of ptosis.[14] He demonstrated an intermittent alternating divergent strabismus with marked limitation of adduction in either eye, a moderate inability to depress either eye and a slight limitation of abduction. A neurological cause could not be identified to explain these oculomotility findings and therefore a myopathic process was

indicated by the author. Although this case details oculomotility defects which are similar to those in our case, the underlying scleroderma is of a systemic origin rather than being purely localised.

The only other reported case, which has the closest relationship to ours, is of an 11-year-old girl with progressive ptosis and a subsequent motility disorder.[15] A slow progression of a right ptosis was observed, which received surgical correction using a right levator resection. The ptosis then re-occurred within 2 years. Ocular motility was initially described as full but at age 16 years diplopia due to a restriction of elevation and abduction of the right eye was reported. A CT scan showed a marked atrophy of the upper levator palpebrae and superior rectus muscle.

Lacey *et al*.[16] carried out a literary review and similarly documented the four main reported cases of LS causing extraocular restriction.[10,13–15] They also carried out a major review in 1999 looking at the clinical records of patients to find the most common causes of muscle disease excluding thyroid orbitopathy. They saw 1750 cases of thyroid eye disease and 99 cases of muscle disease. LS was the cause of chronic extraocular muscle restriction and scarring for 3 of their patients. All patients were female and had a long history cosmetic disfigurement with 'en coup de sabre' and progressive enophthalmos with restriction of ocular motility, particularly in elevation. One example was a 57-year-old woman who presented with progressive, painless enophthalmos of her right eye. An 'en coup de sabre' scar was apparent on her scalp (left), and she had 8 mm of right enophthalmos with restriction of ductions in all directions. A CT scan showed significant enophthalmos, and mild thickening of the right horizontal recti (right) was noted on multiple layers of scanning. Taking into account all the previous relevant articles, ours is the eighth reported case of scleroderma 'en coup de sabre' with associated oculomotility disturbances.[10,13–16]

Finally, an interesting series of editorial reviews looks at the disease in a unique way. Wood[17] published several pieces in successive issues of the *Scleroderma Voice* magazine, looking at physical and functional changes, her own personal experience and what the disease involves. The articles provide readers with an understanding and appreciation of the author's personal symptoms and emotions. She describes many and varied ocular changes including reduced vision, central corneal thickening, Horner's syndrome, mydriasis, heterochromia, or uveitis. But again, even in this personal account motility changes are not described.

These cases highlight the need for clinicians to exclude 'en coup de sabre' as a diagnosis in this age group for patients with progressive ptosis and reducing levator function, or if ptosis re-occurs after correction. This is especially important when there are accompanying skin lesions or motility disturbances. LS can affect children and adults with a multitude of ocular symptoms which can often be progressive; therefore observation after initial presentation should always be advised. Early detection allows early attempts at treatment and other supportive measures. If a diagnosis is known, it is important to accept that surgical intervention may fail and even lead to corneal exposure if eyelid closure is compromised and muscle restrictions subsequently develop.

There is still no common treatment for the ocular complications of 'en coup de sabre'. Each patient has to be reviewed and treated on an individual basis.

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