En Coup De Sabre of The Cheek—A Case Report and Literature Review

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Abstract
The term “en coup de sabre” is used for linear scleroderma located in the temporoparietal region that produces sharp bordered furrowing with alopecia when extending to the scalp. Although it is a benign disease, the resultant marked deformity often necessitates treatment. While in some cases atrophy is confined to the skin and subcutaneous tissue, cases with underlying muscle and bone involvement have also been reported. Symptoms and clinical findings in linear scleroderma of the face show variations. We present a case of “en coup de sabre” linear scleroderma of the cheek affecting only the skin and subcutaneous fat tissue without any accompanying disorder.

Keywords:
en coup de sabre, linear scleroderma, morphea

Introduction
The term scleroderma, which means “hard skin”, is a rare disorder of unknown etiology in which increased collagen deposition occurs and results in dermal thickening. The involvement may be diffuse (systemic sclerosis) or localized to the skin (localized scleroderma) (1).

Linear scleroderma is a variant of localized scleroderma that primarily affects the pediatric population, with 67% of patients given a diagnosis before 18 years of age (2). These lesions are predominantly unilateral and the term “en coup de sabre” is used when the lesions are seen on the scalp or the face. These lesions produce a marked depression on the affected site that resembles the stroke from a dueling sword (3). The lesions described in the literature are mostly in the frontal region and may be accompanied by seizures, dental abnormalities, and ocular muscle dysfunction. We present a case of “en coup de sabre” of the cheek affecting only the skin and subcutaneous tissue without any accompanying disorder.

Case Report
A 14–year–old female patient presented with a painless depression on the right side of her face, which she noticed 4 years earlier (Fig. 1). The lesion had progressed for 3 years and remained stable for the last year. Her childhood health and development had been uneventful and there was no history of trauma or any infections. She sought a medical opinion about the scar on her face and was given topical corticosteroid application, which did not provide relief. Physical examination revealed a depression on the right side of the face extending

![Fig. 1. Photograph of the patient showing the depressed area on the right side of the face.](image-url)
obliquely downwards from the inner canthus of the eye to the corner of the mouth. The depressed area was darker in color than the adjacent normal skin and extended downward in a linear fashion, with its greatest width being approximately 0.5 cm. There was mild facial asymmetry as a result of the depressed area on the right side of the face. The eye movements were in normal range without any visual impairment. Facial electromyography examination performed for neurogenic and myogenic processes revealed normal findings.

The paranasal sinus view (Fig. 2) radiograph revealed hypoplastic frontal sinuses and mild hypoplasia of the right maxillary sinus with mucosal thickening. Hypoplasia of the right maxillary sinus was also revealed in the orthopantomograph. Although a computed tomography scan showed normal findings, T1-weighted axial sections of magnetic resonance imaging of the maxillofacial region revealed hypoplasia of the right maxillary sinus with associated hypoplasia of the overlying soft tissues (Fig. 3). In addition, frontal bones showed decreased pneumatization and the muscles of the face were normal in bulk.

An incisional biopsy was performed on the cheek where the depression was most prominent. Microscopic examination revealed thinning of the epidermis with homogeneous collagen deposition in the dermis. Decreased vascularity with perivascular and periannexal inflammatory infiltrate was observed. Collagen bands showed penetration into the underlying subcutaneous fat tissue. Serological examination results for anti-nuclear antibodies were within normal limits (Fig. 4).

Fig. 2. Paranasal sinus radiograph showing hypoplastic frontal sinuses and mild hypoplasia of the right maxillary sinus (arrow).

Fig. 3. T1-weighted axial section magnetic resonance image of the maxillofacial region showing hypoplasia of the right maxillary sinus with associated hypoplasia of the overlying soft tissues.

Fig. 4. Microscopic section with 40x resolution showing thinning of the epidermis with homogeneous collagen deposition in the dermis and decreased vascularity with perivascular inflammatory cell infiltrate (H and E staining).
The clinical presentation of the lesion with histological findings led us to the diagnosis of linear scleroderma with an “en coup de sabre” presentation. Our patient’s lesion was advanced and hence no pharmacological treatment was instituted. Because the disease is self-limiting, the patient was kept on follow-up and no further atrophy was noticed over the next 6 months. Furthermore, the mild facial asymmetry caused by the hypoplasia of the right maxillary sinus was planned to be corrected by restoring the facial contour with a dermofat graft in case the lesion worsened at follow-up.

Discussion

Linear scleroderma, a variant of localized scleroderma, at the outset belongs to a clinically heterogeneous group of disorders of unknown etiology in which increased collagen deposition occurs, resulting in dermal thickening (4). Localized scleroderma can be divided into five types: plaque morphea, generalized morphea, bullous morphea, linear morphea, and deep morphea (5). There have been several cases reported in the literature that are akin with one of the aforementioned types with or without variations, and our case diagnosed as linear morphea adds to the previously described cases in literature (6).

Linear scleroderma begins with the insidious onset of thickening in the subcutaneous tissues, gradually progressing in a linear pattern. A review of the literature has revealed some case reports suggesting that lesions of linear morphea follow Blaschko’s lines (7, 8). Our case was also found to follow Blaschko’s lines, agreeing with cases described by Feijimoto and Soma (8) and more recently by McKene and Benton (9).

The etiopathogenesis of linear scleroderma remains obscure. Trauma has frequently been implicated with the onset of localized scleroderma (10). Environmental factors like chemicals, toxins, and drugs, including bleomycin and tryptophan, have been associated with systemic sclerosis but are far from common as the etiologic factors in linear scleroderma. Local steroid injections that cause reversible localized scleroderma-like lesions have been described (11). Increased attention has been focused on the role of mast cells in fibrotic disorders. This role has been supported by the fact that increased numbers of mast cells have been found in skin lesions in the inflammatory phase of scleroderma (12). Linear scleroderma has also been found to be associated with infections with *Borrelia burgdorferi* in patients living in certain endemic areas. Such association has been found more often in people living in endemic areas and in patients with progressive facial hemiatrophy (Parry–Romberg syndrome) (13). However, the relationship between *Borrelia burgdorferi* infection and patients with linear scleroderma or progressive facial hemiatrophy has not been confirmed (14).

The linear scleroderma “en coup de sabre” lesions start with an early inflammatory stage and hypere mia of the skin, followed by fibrosis, sclerosis, and finally atrophy. The disease may show a wide variation in its clinical presentation; various clinical abnormalities have been reported in patients with “en coup de sabre” lesions including neurological manifestations, dental abnormalities, and ocular muscle dysfunctions (15–19). Among other ocular manifestations, ocular motility disorders and diplopias have been reported in a number of cases of linear scleroderma (19). Intracerebral lesions such as inflammation, calcifications, and demyelination have been reported in a number of cases of linear scleroderma leading to epilepsy and functional motor defects (20). A review of the literature has revealed that “en coup de sabre” of the cheek presenting as a lesion with no other systemic manifestations is exceedingly rare. The first case of such an appearance on the cheek was reported by Demir et al. in 2003 (21). Our case also gave a clinical presentation of “en coup de sabre” of the cheek with no systemic manifestations.

Linear scleroderma “en coup de sabre” has often been linked with progressive facial hemiatrophy (Parry–Romberg syndrome) in literature and many authors believe that facial hemiatrophy is an intensified form of linear scleroderma (22). However, some authors consider these as two different disease
entities. A review of the literature suggests that the histology of facial hemiatrophy is different from that of linear scleroderma. In linear scleroderma, the primary pathology is in the dermis, while in facial hemiatrophy the subcutaneous tissue is primarily involved (23). Also, in linear scleroderma “en coup de sabre”, inflammatory cell infiltrate around the blood vessels of surfaces and the deep plexes of skin can be observed, which is not seen in cases of facial hemiatrophy (24). The histopathology findings of our case showed perivascular and periannexal inflammatory cell infiltrate.

The histopathology findings of scleroderma include fibrosis with an increased number of fibroblasts and deposition of collagen fibers. The evolution of the sclerodermatous lesion appears to occur in three stages. The first phase is vascular, with the vacuolization and destruction of endothelial cells and the duplication of the basal lamina (6). The second phase is inflammatory, with an influx of lymphocytes, macrophages, mast cells, plasma cells, and eosinophils that interact among themselves and with fibroblasts to increase collagen formation. The third phase involves the proliferation of fibroblasts with an increased deposition of collagen and some perivascular and periadnexal inflammatory infiltrate. However, in the advanced stages, when the skin is hard and indurated, the epidermis is thin and atrophic, the dermis is replaced by dense collagen, and there is atrophy of skin adnexa. The fibrosis extends into subcutaneous fat, giving the impression of widening of the dermis. Fibrosis may also extend into skeletal muscle, with resultant atrophy. The histopathology findings in the biopsy specimen of our patient revealed features akin to those of the third phase of the histopathology with thinning of the epidermis, confirming an advanced lesion of linear morphea (6).

Laboratory tests for localized scleroderma are not diagnostic (25). It has been demonstrated that serological abnormalities have not always been detected in linear scleroderma. In previous studies, anti-nuclear antibodies, including the antibody to Scl-70 antigen, antibody to centromeres, and anti-nucleolar antibodies, were not detected in patients with localized scleroderma (26). Also, anti-topoisomerase II and anti-single-stranded DNA antibodies have been demonstrated in patients with a generalized form of morphea rather than in patients with a more limited cutaneous form of scleroderma (27). Although immune activation is clearly important for the pathogenesis of both localized and systemic sclerosis, the presence of these disparities clearly has some relevance to the differing clinical presentations. Likewise, our patient, presenting no systemic manifestations, was also found to have negative anti-nuclear antibody test results.

Linear scleroderma is a self-limiting disease which, when it occurs in pediatric patients, usually regresses with age. However, cases with underlying muscle and bone involvement causing resultant marked deformity necessitate treatment. When the lesion is active and is rapidly increasing in size, i.e., in the early phases of development, drug therapy is indicated. Treatment should be decided according to the severity and extent of lesions. Limited lesions may be treated with local steroids. Topical steroid therapy should be used for short periods on mildly involved areas (28). Systemic treatment with methotrexate should be considered in extensive and linear forms when there is a risk of functional or esthetic complications. Several other drugs used with varied effects are antimalarials, potassium para aminobenzoate, vitamin E, salazopyrine, etrinate, phenytin, and local and systemic corticosteroids (29, 30). Other drugs like colchicine, cyclosporine A, and interferon gamma have also been used because skin softening is their major therapeutic advantage.

The most commonly used drug on active lesions is penicillamine. The therapeutic effect of penicillamine is caused from its action of inhibiting the intermolecular and intramolecular cross-linking of collagen fibers. This results in a decrease in the formation of collagen fibers but has no effect on preformed collagen fibers (31).

Topical application of calcitropin, a synthetic analogue of vitamin D, exerts its effect on early lesions by inhibiting the synthesis of inflammatory
mediators such as interleukin-2 and by decreasing the collagen synthesis (32).

The introduction of phototherapy and phototherapic for sclerosing skin diseases has considerably enriched the therapeutic panel and proven useful in a number of sclerosing skin diseases including linear scleroderma. Two phototherapeutic modalities are used for the treatment of linear scleroderma: long-wave ultraviolet A (UVA) and psoralen plus ultraviolet A (PUVA). The treatment consists of oral methoxsalen (0.4 mg/kg body weight) followed by exposure of the affected part only to ultraviolet-A radiation on a regular schedule of three treatments each week. A brief maintenance treatment, weekly and then biweekly exposures, should be given (33). PUVA therapy suppresses the inflammatory phase of morphea by a cytotoxic effect on the cells responsible for the inflammation or by inhibition of the mediators causing the changes in collagen.

Newer treatment modalities of linear scleroderma include pulsed dye laser, 5% imiquimod cream, and bosentan. Studies have previously shown that improvement of hypertrophic scars and fibrotic skin can be achieved with the use of a 585-nm pulsed dye laser. The mechanism of its effect in this condition remains unknown (34).

In one case report, 5% imiquimod cream, which induces interferon and in turn inhibits TGF-beta, was employed to treat morphea (35). Bosentan, an endothelin receptor antagonist with vasodilative and antifibrotic properties, was successfully used to treat refractory cutaneous ulcerations in pansclerotic morphea (36).

Conclusion

Linear scleroderma with an “en coup de sabre” presentation on the cheek, a disease that primarily affects the pediatric population, is rarely found to occur without any systemic manifestations. Its clinical presentation, as seen in our patient, is characteristic and the histological findings, although not solely diagnostic, give a definite clue towards confirming the diagnosis. Likewise, the diagnosis of our case was made on clinical grounds, with the histological findings confirming our diagnosis. Anti-nuclear antibodies, which are mostly found in more generalized forms, were not found in our case, which had limited cutaneous involvement. This is a self-limiting disease and treatment is advocated depending upon the severity and stage of the lesion.

References