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FRONTAL BONE OSTEOMA AS A CHRONIC BIOMECHANICAL TRIGGER FOR EN COUP DE SABRE

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ABSTRACT
Osteomas and localized morphea are conditions that may be triggered by trauma. This report describes an early adolescent female who presented with localized craniofacial linear morphea, also known as en coup de sabre (ECDS). The lesion developed following blunt force trauma that resulted in a frontal bone osteoma and transient neurological deficits, including a generalized seizure and hemiparesis. Magnetic resonance imaging (MRI) showed no intracranial abnormalities, however, the progressive development of the sclerotic plaque suggested a possible interaction between skeletal injury and autoimmune fibrotic dysregulation. This case highlights the possibility of a Deep Koebner phenomenon in morphea, where the underlying post-traumatic osteoma likely served as a persistent stimulus for overlying cutaneous and subcutaneous fibrosis. The therapeutic challenge posed by the refractory nature of the lesion, which progressed despite treatment with systemic methotrexate, pulse corticosteroids, and targeted 308-nm excimer therapy, emphasizes the need for early and multidisciplinary management of pediatric craniofacial morphea to prevent functional and cosmetic complications.

KEYWORDS: En coup de sabre; Localized scleroderma; Frontal osteoma; Koebner phenomenon; Pediatric morphea.

INTRODUCTION
Morphea or localized scleroderma is a rare autoimmune disease, which primarily affects the skin, and is characterized by an inflammatory stage followed by sclerosis and then later atrophy or a “burnt out stage” [1]. It is characterized by a chronic fibrosing connective tissue reaction driven by dysregulated dermal fibroblast activity. Fibroblasts derived from lesional skin exhibit increased collagen synthesis and reduced collagenase activity, resulting in excessive extracellular matrix deposition [2]. Genetic susceptibility has also been implicated and is associated with HLA-DRB1*04:04 and HLA-B37.Depending on the subtype and localization, nearby structures including fat, muscles, joints, and bones may be affected. Linear subtype scleroderma can affect the face, frontoparietal region is referred to as En coup de sabre. Herein we describe a rare phenomenon of deep Koebner phenomenon in morphea from a post traumatic osteoma [3, 4].

Case Presentation
An early adolescent female child was referred by her pediatrician for evaluation of a progressively enlarging swelling and hyperpigmentation on the right side of the forehead (Figure 1). The patient’s medical history was unremarkable until 6 months prior to presentation, when she sustained a fall from bed. Immediately following trauma, she experienced a transient episode of neurological deficit characterized by stiffening of the left upper lip and weakness of the left upper and lower limbs. This episode lasted approximately 20 minutes and resolved spontaneously. She also experienced a single episode of generalized tonic-clonic seizure.

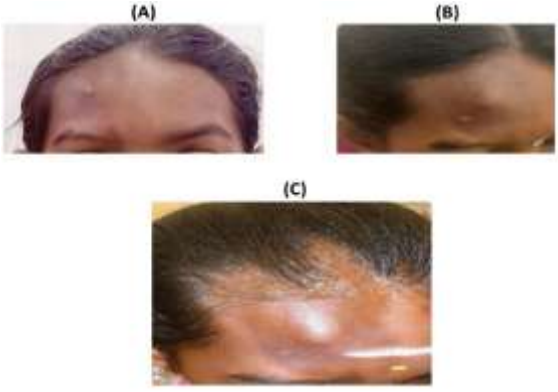

Figure 1. Clinical progression of post-traumatic en coup de sabre morphea overlying a frontal bone osteoma. (A, B) Initial clinical presentation showing a localized firm swelling over the right frontal region with subtle

overlying hyperpigmentation following blunt head trauma. (C) Clinical appearance after 2 years demonstrating progressive enlargement of the frontal swelling with extension of hyperpigmentation toward the medial eyebrow and frontal hairline, consistent with active linear morphea (en coup de sabre), accompanied by localized alopecia and cutaneous sclerosis.

Approximately two months after the initial trauma, the parents noted a firm, non-mobile swelling on the right side of the forehead. One month later, the area surrounding the swelling showed a blackish discoloration, that gradually progressed towards the medial aspect of the right eyebrow, accompanied by localized loss of hair. The patient also had a head injury at age 2 in the same anatomical region which was not associated with neurological sequelae.
Dermatological examination revealed a single, ill-defined swelling measuring 5 x 6 cm on the right side of the forehead. Upon palpation, the mass was found to be bony hard, immobile, mildly tender, and fixed to the underlying skull. Overlying the swelling, there was a patch of ill-defined hyperpigmentation that extended to the medial aspect of the right eyebrow associated with loss of hair. The skin over the hyperpigmented patch was pinchable, although thickened compared to the contralateral side. No focal neurological deficits were present at the time of examination, and the cutaneous assessment did not reveal any other skin lesions or signs of systemic disease.

Investigations
Dermoscopy showed absence of follicular openings with multiple eccrine duct openings and an ill-defined homogeneous white area representing scarring. Punch skin biopsy demonstrated thinned out epidermis with loss of appendages and dermis showed (Figure 2) parallel arrangement of thickened hypo cellular collagen with minimal lymphoplasma cell infiltrates. Fibrosis extended into subcutaneous tissue and underlying skeletal muscle bundles suggestive of Morphea.

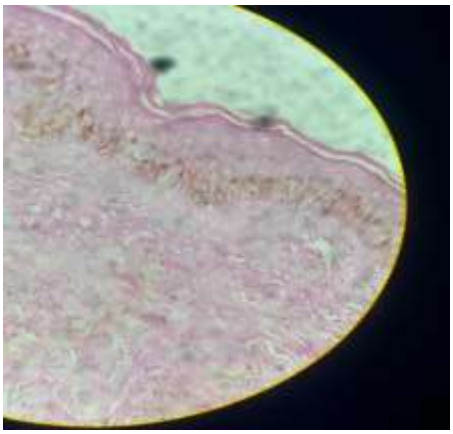

Figure 2. Histopathological features of morphea. Representative hematoxylin and eosin-stained section showing epidermal thinning with dense, hypocellular, thickened collagen bundles arranged in a parallel fashion within the dermis (arrow). Fibrosis extends into the subcutaneous tissue and underlying skeletal muscle with minimal lymphoplasmacytic inflammatory infiltrate and loss of adnexal structures, consistent with deep morphea (localized scleroderma).

Ultrasonographic examination of the swelling and soft tissue demonstrated soft tissue edema with no cystic lesions. Radiological evaluation (Figure 3) of the skull identified a localized radiopaque lesion on the frontal bone suggestive of osteoma. ANA Immunofluorescence was negative. The above findings helped us confirm the diagnosis of Morphea, further MRI brain with venogram and angiogram, and EEG studies were done to investigate the patient's history of seizures and transient hemiparesis, but the results were normal and showed no electrical or parenchymal abnormalities (Figure 4).

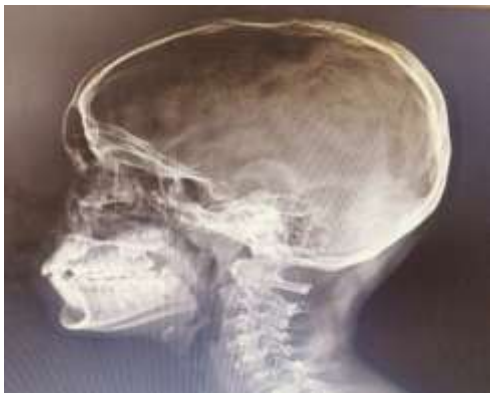

Figure 3. Plain radiograph demonstrating frontal bone osteoma. Lateral skull radiograph showing a well-defined localized radiopaque lesion arising from the right frontal bone (arrow), consistent with a benign osteoma at the site of previous trauma. No associated osteolytic changes or intracranial bony abnormalities are identified.

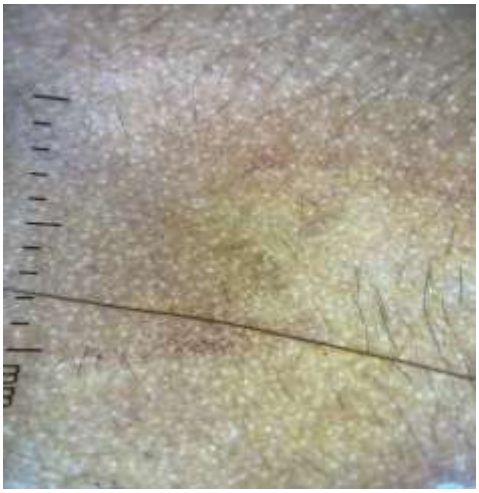

Figure 4. Dermoscopic features of active en coup de sabre morphea. Dermoscopic examination of the affected forehead demonstrating absence of follicular openings, multiple preserved eccrine duct openings, and an ill-defined homogeneous white structureless area corresponding to dermal sclerosis and scarring. These findings are characteristic of localized morphea.

Differential Diagnosis

This clinical presentation of post-traumatic bony mass with overlying progressive sclerosis associated with transient neurological symptoms made us consider Morphea and Langerhan cell histiocytosis as differential diagnosis [5].

Treatment

The patient was started on weekend oral minipulse of betamethasone (2 mg), clobetasol-calcipotriene combination ointment, and tacrolimus ointment 0.1%, in addition to targeted phototherapy with excimer 308 nm. Despite adherence to this multimodal therapy, clinical response was suboptimal, with continued disease activity and progression of lesions toward the frontal hairline. Due to the inadequate therapeutic response, systemic methotrexate was initiated following comprehensive laboratory evaluation.

Outcome and Follow-Up

At present, the patient remains in the active phase of her disease with slow but persistent progression despite systemic therapy, this is consistent with the nature of deep morphea in pediatric age group which may require prolonged treatment before reaching inactive stage. Neurologically her status remains stable with no further seizure activity or focal deficits, and she continues to undergo regular monitoring by a multidisciplinary team including pediatric dermatology, rheumatology, neurology and ophthalmology due to the increased risk of subclinical uveitis. The presence of the osteoma remains a unique clinical variable (Figure 5).\

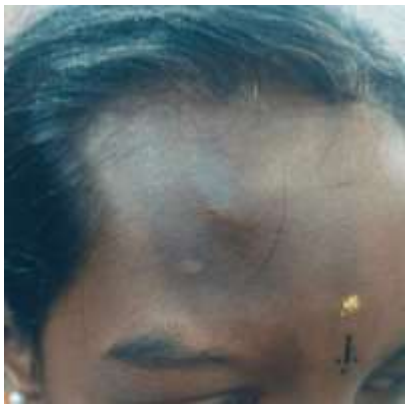

Figure 5. Clinical follow-up after 2.5 years of treatment. Follow-up clinical photographs obtained after approximately 2.5 years of systemic and topical therapy demonstrating persistent frontal bony prominence with residual hyperpigmentation and localized sclerosis. Although neurological symptoms remained stable, cutaneous disease activity persisted with slow progression, reflecting the refractory nature of deep craniofacial morphea associated with the underlying frontal osteoma.

While surgical resection of the osteoma could theoretically remove the chronic mechanical trigger, operative trauma itself may provoke further disease activation through Koebnerization. Therefore, current management focuses on achieving medical control of the inflammatory and fibrotic process before considering any structural interventions.

DISCUSSION

Craniofacial localized morphea can be sub-classified into ECDS which presents as a linear atrophic band on the forehead or scalp, and Parry-Romberg syndrome (PRS), also known as progressive hemifacial atrophy. Both conditions belong to the same clinical spectrum and are frequently associated with neurological and ophthalmological complications, including seizures, headaches, and uveitis [6, 7]. The prevalence of central nervous system (CNS) involvement in craniofacial morphea is reported to be as high as 44% to 50%, often manifesting as "silent" intracranial abnormalities on MRI [6].

The exact pathogenesis of morphea remains incompletely understood but is thought to involve a multifactorial interplay of genetic predisposition, vascular injury, and autoimmune dysregulation. Genetic associations with specific human leukocyte antigen (HLA) alleles, such as DRB1*04:04 and B*37, suggest an underlying susceptibility to aberrant immune responses [8]. Environmental factors, including physical trauma, radiation, and infections can act as precipitating events for fibrotic process. Trauma-induced morphea accounts for approximately 16% of cases, occurring via isomorphic or isotopic responses [9].

The clinical course of this patient illustrates the potential of physical trauma acting as a possible etiology for complex, self-perpetuating autoimmune response. The development of osteoma at the site of impact, followed by the development of a morphea plaque, suggests an integrated mechanical and immune pathology. Traditionally, Koebner phenomenon or isomorphic response describes the appearance of lesions on healthy skin following superficial trauma. However, the concept of the "Deep Koebner" phenomenon extends this reaction to deeper structural injuries. Similar mechanism has been described in psoriatic arthritis, where micro-trauma at entheses triggers localized inflammation and bone remodeling [5, 10]. In our patient, the frontal bone injury resulted in an osteoma, a benign but persistent skeletal irregularity that likely induced a chronic biomechanical stress on the overlying dermis and subcutaneous tissues. Pathogenesis studies in morphea suggest that recurrent tissue damage upregulates damage-associated molecular patterns (DAMPs) and toll-like receptor (TLR) ligands on fibroblasts. This activation enhances pro-fibrotic signaling pathways, leading to excessive collagen deposition [11]. The presence of the osteoma may have established a self-amplifying cycle of fibrogenesis by providing a constant irregular surface for friction and tension.

The patient's transient neurological deficits further support the extracutaneous manifestations of craniofacial morphea. Interestingly, neurological symptoms frequently do not correlate with MRI findings. Many patients with striking intracranial abnormalities remain asymptomatic, while others with normal imaging experience profound symptoms. This neuro-radiological paradox suggests that the neurological manifestations may be due to transient inflammatory vasculitis or localized metabolic changes that are not always captured by standard anatomical imaging. Additionally, the history of head injury at age 2 in the same region also raises the possibility of an isotopic response (Wolf's isotopic response) where a site of prior injury remains immunologically primed, and susceptible to the development of another unrelated skin condition upon subsequent insult [12].

Pediatric morphea is generally more aggressive than its adult counterpart, with a higher frequency of deep tissue involvement and a longer duration of active disease [13]. Histopathological evidence of skeletal muscle involvement in this patient is an indicator of disease severity, as deep variants often result in permanent atrophy and contractures. The continued progression of the lesions despite multi-modal therapy indicates a refractory phenotype, potentially exacerbated by the persistent biomechanical stimulus of the underlying osteoma.

Therapeutic strategies for localized morphea has evolved from conservative topical approaches to aggressive systemic immunomodulation, particularly for linear and deep subtypes where the risk of deformity is high. Initially, the patient was treated with high-potency topical corticosteroids, tacrolimus 0.1% ointment and calcipotriene. Calcineurin inhibitors such as tacrolimus act by inhibiting T-lymphocyte activation and reducing local cytokine production, while vitamin D analogues modulate the fibroblast proliferation and collagen synthesis hence are effective in mild, superficial plaques(14). However, due to the depth of the lesion and the progressive nature of the disease, systemic therapy was required. Methotrexate (MTX) remains the gold standard for juvenile linear morphea, typically administered at 15 mg/m2 weekly [14]. MTX acts as a folic acid antagonist, reducing the proliferation of inflammatory cells and inhibiting pro-fibrotic pathways. To induce rapid remission, MTX is frequently paired with pulse steroids. This patient received weekend oral betamethasone sodium to minimize daily steroid side effects while maintaining an anti-inflammatory state during the active phase of the disease.

In addition to the topical and systemic therapy, targeted phototherapy with an excimer laser (308 nm) was initiated due to ongoing progression. The 308nm wavelength is highly effective for localized sclerotic conditions because it penetrates the dermis to induce apoptosis of T-cells and downregulate pro-inflammatory cytokines like IL-6 and TNF-alpha [15].

Given the continued extension towards the hairline, the patient may be a candidate for biological therapy. Tocilizumab, a humanized monoclonal antibody against the IL-6 receptor, has shown significant efficacy in refractory juvenile morphea by interrupting the IL-6 driven fibrotic cascade, while other emerging options include Abatacept (CTLA-4 fusion protein) and JAK inhibitors [16].

CONCLUSION

In conclusion, this case highlights the complex interplay between trauma, structural abnormality induced by osteoma and immune dysregulation in the pathogenesis of craniofacial morphea. It emphasizes the role of deep tissue injury and biomechanical stressors in refractory disease and illustrates the importance of recognizing extracutaneous manifestations and neuro-cutaneous dissociation. Early aggressive systemic therapy and multidisciplinary monitoring are essential in pediatric patients to prevent deformities. Furthermore, this coexistence warrants further investigation into the role of deep Koebner phenomenon in sclerosing disorders.

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