Frontal bone loss following coronal brow lift: A mimicker of head variant linear morphea

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INTRODUCTION

Linear morphea is a rare sclerosing disease involving the presence of skin tightening and increased collagen, with potential atrophy of underlying structures. The head variant of linear morphea, "*en coup de sabre*" (ECDS), presents as unilateral thickening of skin on the paramedian or frontoparietal forehead with or without atrophy of underlying structures, which can result in functional neurologic, ocular, and oral symptoms.[1,2] This case highlights a complication of coronal brow lift surgery that can mimic ECDS.

Abbreviation used:

ECDS: *en coup de sabre*

CASE REPORT

A woman in her 70s with a history of osteopenia presented with bilateral depressions of the frontal scalp and forehead that progressed over 3 years and were mildly pruritic without associated neurologic symptoms. She had a remote history of a coronal brow lift surgery more than 20 years prior and denied more recent local procedures or trauma. The patient has no history of prior bisphosphonate use. Physical examination demonstrated a symmetric, arcuate depression along the frontal hairline and depression of the central forehead; there was a subtle ivory-white change of the overlying skin without induration and hair thinning of the frontal scalp with remaining overlying hair preserved (Fig 1). Ocular, oral, and other facial structures were symmetric and uninvolved. Facial muscle strength was normal and symmetric bilaterally.

Radiological studies, including radiographs, computed tomography, and magnetic resonance imaging, revealed concave deformities of the calvarium with associated atrophy of scalp soft tissue and severe thinning of the frontal bone without underlying brain pathology or evidence of malignancy, infection, or metabolic bone disease (Figs 2 and 3). A nuclear medicine scan showed no significant increase in bone turnover. Skin biopsy findings demonstrated nonspecific features, including thinning of the dermis, subtle sclerosis of the reticular dermis, and sparse inflammatory infiltrate; overall, these findings were considered potentially consistent with, though not significant enough to be diagnostic of, morphea. Additional inflammatory, metabolic, and endocrine work-ups,

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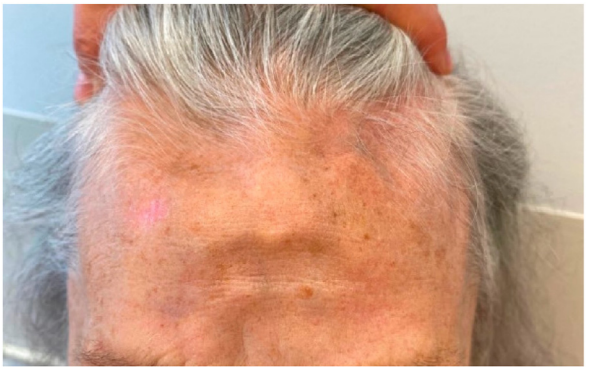

Fig 1. Frontal bone loss: initial physical examination of a patient with depression of the central forehead.

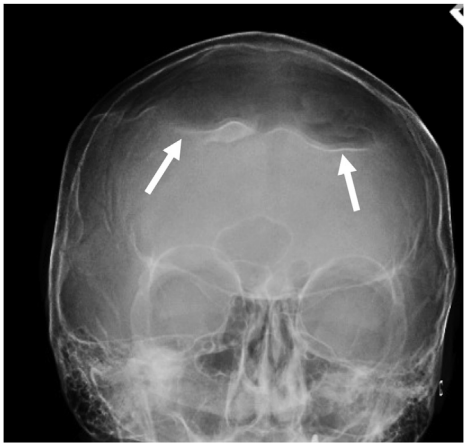

Fig 2. Frontal bone loss: skull radiograph revealing bilateral depression of the frontal bones and atrophy of the scalp soft tissue about the vertex of the frontal bone.

including C-reactive protein, erythrocyte sedimentation rate, parathyroid hormone, and alkaline phosphatase, were unremarkable. Bone turnover studies, such as serum cross-linked C-telopeptide of type I collagen, amino-terminal propeptide of type 1 procollagen, and bone-specific alkaline phosphatase, were unrevealing. Infectious work-ups, including tuberculosis and coccidioidomycosis testing, were negative.

Given concern for an unusual presentation of ECDS, the patient was treated with mycophenolate mofetil up to 1500 mg twice daily for 1 year. However, despite treatment, her disease did not improve and continued to gradually progress in a horizontal configuration, in contrast to the vertical orientation of classic ECDS (Fig 4). Multiple repeat radiographic studies showed stability of the bilateral frontal bone depressions without involvement of the brain. Following multidisciplinary evaluation with rheumatology, endocrinology, neurology, neurosurgery, and radiology, mycophenolate mofetil was discontinued as the favored diagnosis was frontal bone loss secondary to coronal brow lift surgery, a rare complication previously reported in the literature.[3] Following the discontinuation of immunosuppressive therapy, the patient was started on bisphosphonate therapy. After 2 months, the patient reported significant improvement in her itching. Overall, the patient's focal frontal bone loss has remained on a stable trajectory, and she is being monitored for the need for protective plating of the calvarium.

DISCUSSION

We present this case as a rare complication of coronal brow lift surgery that may mimic ECDS and should be considered in the differential diagnosis, particularly in cases with atypical morphology and a history of cosmetic surgery. Although ECDS classically presents as a unilateral, indurated sclerotic plaque of the paramedian or frontoparietal scalp in a vertical orientation, this patient presented with horizontal indentations, the most prominent of which were at the suture lines from her prior coronal brow lift surgery. In patients with an atypical presentation of suspected ECDS, it may be helpful for providers to elicit a history of cosmetic surgery, in addition to evaluation and work-up for other similarly rare but more concerning etiologies. Reassuringly, in this patient, her work-up was unrevealing, and specifically, the absence of facial atrophy or brain abnormalities such as white matter disease, microhemorrhage, or hemispheric atrophy was not consistent with Parry-Romberg syndrome, and the absence of pain and complete bony resorption was not consistent with Gorham-Stout disease.

There has been only 1 previously reported case with nearly identical features with frontal bone indentations 3 years following coronal brow lift surgery, without evidence of ECDS, Parry-Romberg syndrome, or other metabolic, endocrine, neurologic, or infectious processes.[3] The pathogenesis of the associated bone loss is postulated to be secondary to surgical disruption of vascular and lymphatic circulation, leading to impaired tissue perfusion and localized bone resorption.[3] Although our case presented after a notable latency period of over 20 years, there have been reports of delayed onset of bone loss following other forms of facial cosmetic surgery. For example, patients undergoing silicone rubber chin implants were found to have concave erosions of the mandible underlying their implants with a delayed latency of up to 25 years.[4,5]

Overall, our case highlights the rare complication of focal frontal bone loss following coronal brow lift

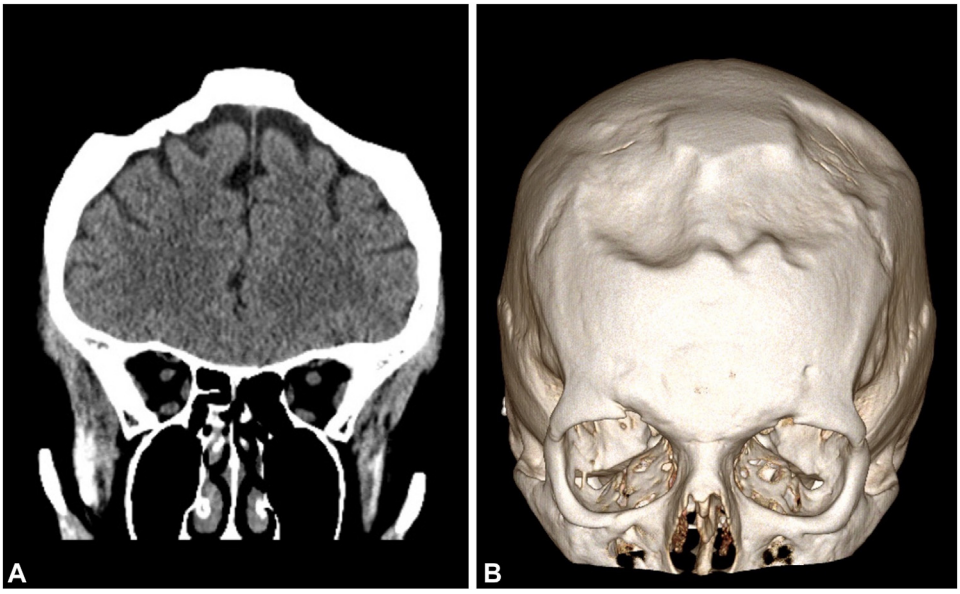

Fig 3. Frontal bone loss. **A,** Coronal view of the computed tomography (CT) brain demonstrating thinning of bilateral frontal bones along with bilateral scalp atrophy about the vertex. **B,** Three-dimensional reconstruction of the CT brain without contrast reveals a smoothly marginated concave deformity of the bilateral frontal calvarium.

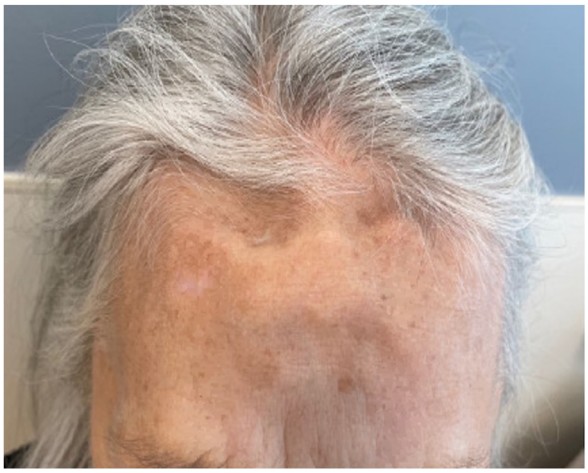

Fig 4. Frontal bone loss: Progression of bilateral depressions of frontal bone loss after 1 year of therapy on mycophenolate mofetil.

surgery that should be included in the differential diagnosis for clinical presentations concerning for atypical ECDS. These atypical cases benefit from multidisciplinary evaluation and work-up, which can limit the unnecessary use of systemic therapies. Further collaborative investigation is needed to clarify the pathophysiology and treatment of this condition.

Conflicts of interest

Dr Hsiao serves in a volunteer capacity on the Medical Registry Advisory Board of the International Fibrodysplasia Ossificans Progressiva Association and on the Fibrous Dysplasia Foundation Medical Advisory Board, is a member of the International Clinical Council on FOP, and receives clinical trials support from Clementia Pharmaceuticals, an Ipsen Company; and Ultragenyx, Inc. Dr Haemel is a consultant to Guidepoint LLC and to CSL Behring. Author Kazmi and Drs Zhang, Gross, Grouse, Link, Tetzlaff, and Theodosopoulos have no conflicts of interest to declare.

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