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Neuroimaging and clinical findings in a case of linear scleroderma en coup de sabre

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ABSTRACT

Linear scleroderma "en coup de sabre" is a subset of localized scleroderma with band-like sclerotic lesions typically involving the frontoparietal regions of the scalp. En coup de sabre and Parry–Romberg syndrome are variants of linear morphea on the head and neck that can be associated with neurologic manifestations. On imaging, patients may have lesions in the cerebrum ipsilateral to the scalp abnormality. We present a case of an 8-year-old girl with a left frontoparietal "en coup de sabre" scalp lesion and describe the neuroimaging findings of frontoparietal white matter lesion discovered incidentally on routine magnetic resonance imaging. The patient had no neurologic symptoms given the lesion identified.

Introduction

Linear scleroderma "en coup de sabre" (ECDS) is a rare subset of localized scleroderma. Localized scleroderma is a rare disease seen in both adults and children. Most pediatric patients have the linear subtype, which can extend deeply into the subcutaneous tissue, muscle, and bone. Affected individuals typically have a characteristic atrophic skin lesion involving the frontoparietal scalp. The disease usually has a benign course and has been distinguished in the past from systemic scleroderma by lack of significant internal organ involvement [1]. Linear scleroderma on the head and neck, called en coup de sabre, and Parry–Romberg syndrome (PRS), also called progressive hemifacial atrophy, are felt to be related variants within the scleroderma spectrum of disease [1]. On rare occasion evidence of organ involvement in linear scleroderma can be manifested as involvement of the rheumatologic, neurologic, and ophthalmologic systems [2]. Additionally, rare neurologic symptoms can be seen associated with linear scleroderma. The most common neurologic symptom is epilepsy, but other neurologic deficits like movement disorders or behavioral changes have been reported [2]. The presence of neurologic symptoms often heralds the existence of an intracranial abnormality. Both ECDS and PRS may be associated with cerebral inflammation and neurologic abnormalities. A variety of neurologic symptoms have been reported, most commonly seizures and headaches [2]. In addition, magnetic resonance imaging (MRI) can reveal calvarial and intracranial abnormalities, even in asymptomatic patients [3]. Cranial MRI findings seen in this group of patients commonly include: focal brain atrophy, calcifications and T2 hyperintense white matter lesions that may demonstrate contrast enhancement [2,3]. Characteristically, white matter lesions and calcifications are found in the cerebral

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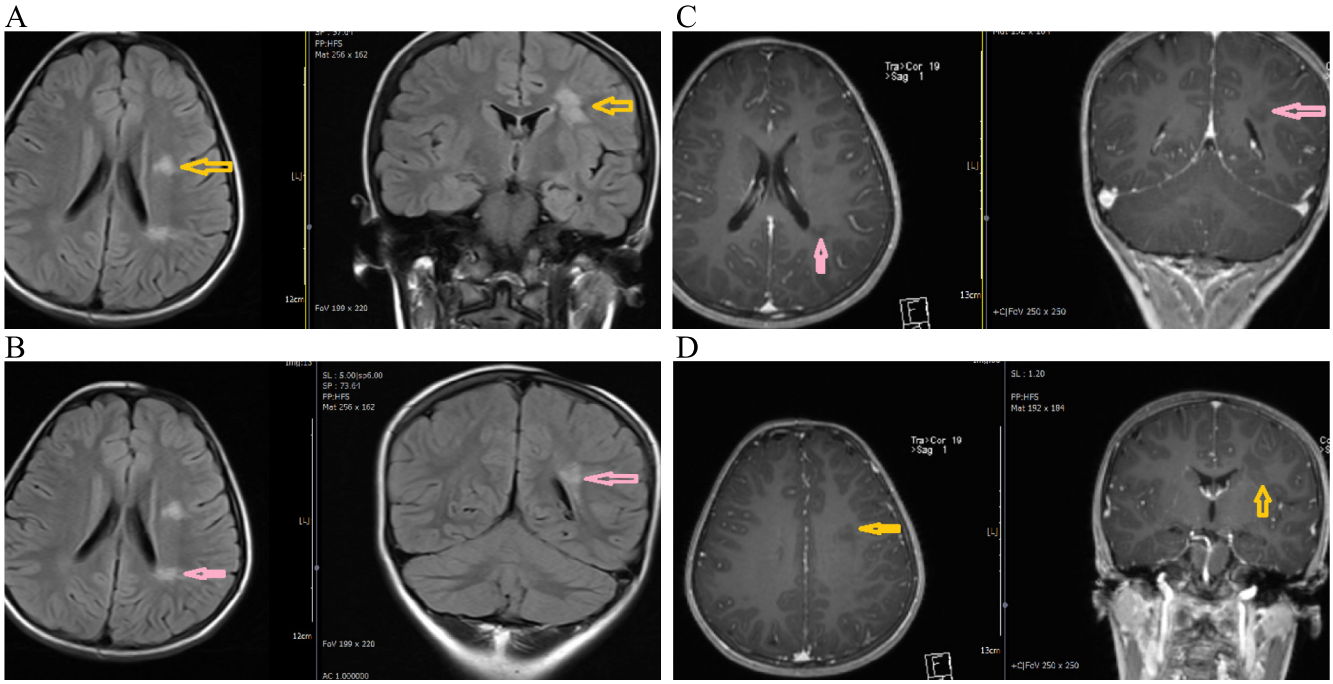

Fig. 1 – (A) Left: axial T2 fluid attenuation inversion recovery (FLAIR) image demonstrates frontal periventricular subcortical white matter lesion (yellow arrow). Right: coronal T2 FLAIR image demonstrates periventricular subcortical white matter lesion (yellow arrow). (B) Left: axial T2 FLAIR image demonstrates parietal periatrial subcortical white matter lesion (pink arrow). Right: coronal T2 FLAIR image demonstrates parietal periatrial subcortical white matter lesion (pink arrow). (C and D) Left upper, right upper: axial and coronal postcontrast T1 weighted multiplanar reformat imaging, respectively. Frontal periventricular subcortical white matter lesion shows mild enhancement (yellow arrow). Left lower, right lower: axial and coronal postcontrast T1 weighted multiplanar reformat imaging, respectively. Parietal periatrial subcortical white matter lesion shows mild enhancement (pink arrow).

hemisphere ipsilateral to the skin abnormality. Early recognition of neurologic involvement in these children is important so that appropriate treatment with systemic medications may be initiated. This report describes the case of a young girl with linear scleroderma ECDS, who was found to have a frontoparietal white matter lesion on incidental MRI. In this paper, clinical presentation of linear scleroderma ECDS and its neurological involvement are described.

Case report

An 8-year-old girl was referred for routine brain MRI by the department of neuroradiology after presenting for consultation in the setting of her primary illness, linear scleroderma ECDS, affecting her left frontoparietal scalp. The patient was initially diagnosed with linear scleroderma at that time on the basis of physical exam findings. At the time of diagnosis, as well as at the time of the first MRI, the patient had no neurologic deficits or neurologic symptoms. On physical examination, the patient had a band-like scalp lesion in the left upper frontoparietal region consisting of a 7.5 × 2.5 cm atrophic, shiny, pink-white plaque with alopecia and hyperkeratosis. A brain MRI was performed using a 3-T MR scanner (MAGNETOM Verio, Siemens Healthcare, Erlangen, Germany). At the time of diagnosis, imaging demonstrated a 20 × 12 mm T2 hyperintense lesion in the periventricular subcortical white matter of the left frontal lobe and a 20 × 10 mm T2 hyperintense lesion in the periatrial subcortical white matter (Figs. 1A and B). On post contrast images, there was mild enhancement in the lesion (Figs. 1C and D). Axial and coronal T2 fluid attenuation inversion recovery, T1 weighted images demonstrated a focal area of scalp thinning within the left frontoparietal region with associated flattening and thinning of the underlying frontal and parietal bones. There was no evidence of intracranial calcifications (Figs. 2A and B).

Discussion

Linear scleroderma ECDS and PRS is a rare subset of localized scleroderma, a distinct and separate disease entity from systemic scleroderma [1]. Localized scleroderma is also referred to as morphea, and is differentiated from systemic sclerosis by the absence of sclerodactyly, Raynaud's phenomenon, capillaroscopic abnormalities, and organ involvement. Localized scleroderma is a fibrosing condition characterized by thickening and hardening of the skin as a result of increased collagen production, with involvement of the subcutaneous tissue and underlying bone. Five clinical subtypes of localized