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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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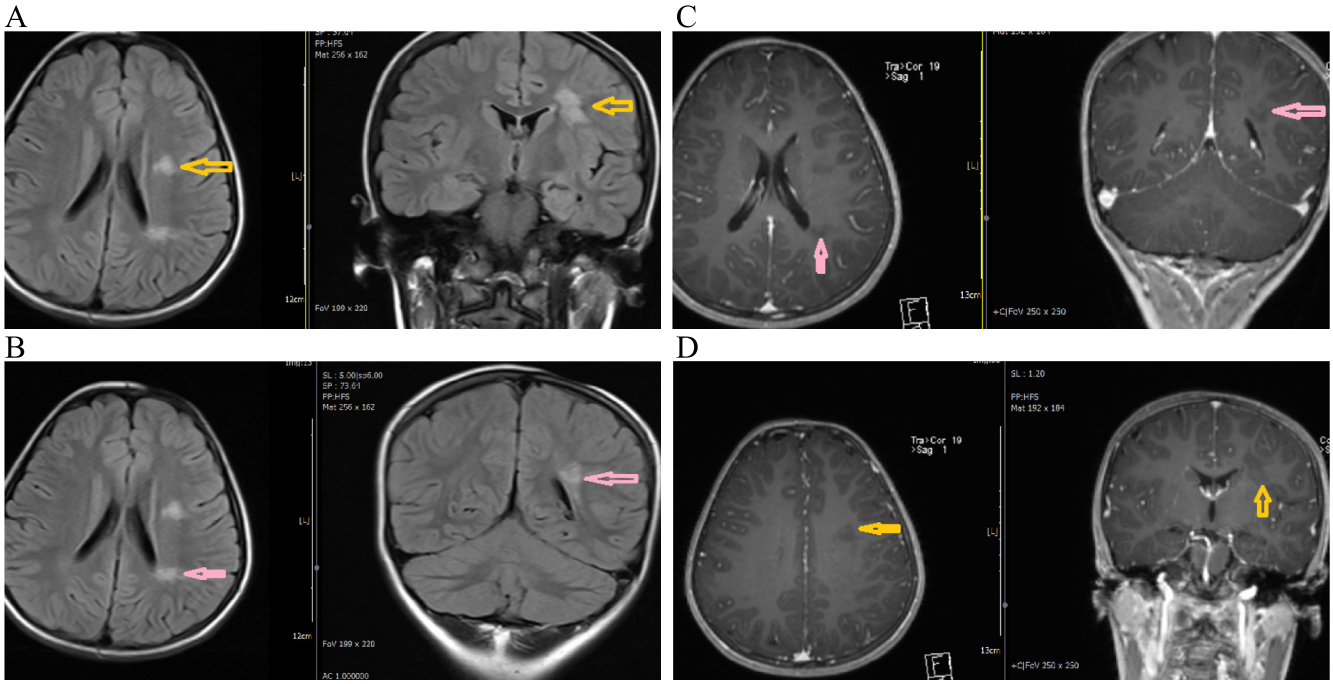

Fig. 1 – (A) Left: axial T2 fluid attenuation inversion recovery (FLAIR) image demonstrates frontal periventricular subcortical white matter lesion (yellow arrow). Right: coronal T2 FLAIR image demonstrates periventricular subcortical white matter lesion (yellow arrow). (B) Left: axial T2 FLAIR image 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

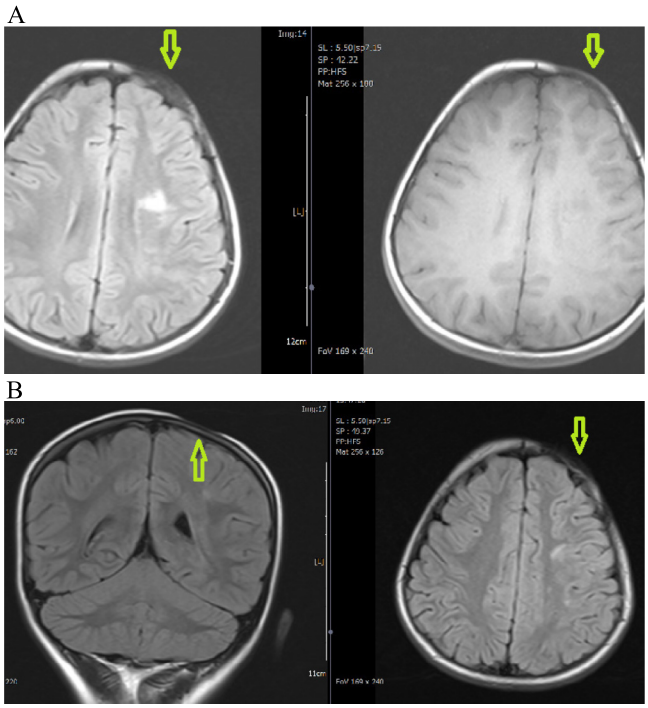

Fig. 2 – (A and B) Left upper; coronal T2 FLAIR image, right upper; axial T2 FLAIR image, left lower; axial T2 FLAIR image, right lower; axial T1 weighted image, respectively. Images demonstrate a focal area of scalp thinning within the left frontoparietal region with associated flattening and thinning of the underlying frontal and parietal bones (yellow arrow).

scleroderma have been suggested: circumscribed, linear, generalized, pansclerotic, and mixed [1]. Linear scleroderma is the most common subtype in children and has three clinical variants: linear limb involvement, ECDS, and progressive hemifacial atrophy (PRS) [1,4]. Underlying tissue atrophy is commonly present in these three variants. ECDS is defined by a unilateral band of sclerotic skin lesions in the frontoparietal area of the head resembling a deep saber wound, hence the name "en coup de sabre" (meaning, the saber cut in French). ECDS can be associated with ipsilateral underlying intracranial lesions and ocular involvement [1]. Progressive hemifacial atrophy (PHA) consists of atrophy which extends below the skin and subcutaneous tissues, and often involves underlying muscle and bone [2]. Several authors have postulated that ECDS and PHA may be clinical variants of the same disease [1–4]. These 2 entities can coexist and share similarities including a comparable age of onset, female predominance, identical neurologic and ophthalmologic complications, and the same neuroimaging characteristics. Both diseases may respond to immunosuppressive treatment [2]. The incidence of localized scleroderma has been reported as 0.4 to 2.7 per 100,000 people [1], with a female predominance, and increased prevalence in the white population [1]. The skin lesions of localized scleroderma are characteristic clinical findings that undergo an initial inflammatory stage of erythematous, dusky, violaceous patches, or plaques. Later, the center of the lesion becomes white and sclerotic, with a surrounding outer "violaceous ring". Once the active phase resolves, the skin lesion turns into a near completely white sclerotic plaque with subsequent post-inflammatory hyperpigmentation. Excessive collagen deposition destroys hair follicles and adnexal structures, resulting in hairless, anhidrotic plaques [1]. Aside from alopecia in a band-like fashion in the frontoparietal scalp and forehead, cutaneous manifestations may extend to the nose, cheek, chin, and neck. Facial atrophy occurs if the underlying muscle, cartilage, and bone are involved. These clinical findings can overlap or coexist in ECDS and PHA making it difficult to classify them as separate entities. In most reported cases of linear scleroderma in the literature, neurologic symptoms are usually preceded by the skin manifestations; however, neurologic symptoms can sometimes appear first. The range of neurologic symptoms associated with craniofacial scleroderma is variable. They include seizures, recent onset headaches, focal neurologic deficits and movement disorders which can be secondary to brain lesions, trigeminal neuralgia, masticatory spasms, mimics of hemiplegic migraines, behavioral changes, or progressive intellectual deterioration due to cerebral hemiatrophy either with or without focal seizures [2,4]. Epilepsy remains, however, the most common neurologic symptom associated with linear scleroderma [2]. Uncommonly, asymptomatic patients may be found to have abnormal neuroimaging studies. Classic central nervous system findings on computed tomography and MRI include brain parenchyma atrophy, white matter lesions, and focal subcortical calcifications. Parenchyma and leptomeningeal enhancement may also be seen [2,3]. Neuroimaging findings are typically ipsilateral to the skin lesions in the cerebral hemisphere. Rarely, contralateral and infratentorial involvement have been described [2,5]. Calcifications typically involve the basal ganglia, thalami, and dentate nuclei, but can also be found in the subcortical white matter [2,6]. MRI usually exhibits T2 hyperintensities, mostly in the subcortical white matter, but also in the corpus callosum, deep grey nuclei, and brain stem. Cerebral atrophy is generally focal and subtle, characterized by blurring of the gray-white interface, cortical thickening, and abnormal gyral pattern [2,3]. Rarely, hippocampal atrophy has been reported [6]. The diagnosis of linear scleroderma is made based on the clinical characteristics of the cutaneous and soft tissue findings. There are no laboratory tests diagnostic for linear scleroderma, although, patients may be positive for antinuclear antibodies, antisingle-stranded DNA antibodies and rheumatoid factor [2,4]. The etiology of linear scleroderma is not well-understood; however, there is evidence suggesting an autoimmune origin of this disease [2]. Early in the disease course, damage to the endothelial cells results in increased vascular permeability with mononuclear cell infiltration, perivascular inflammatory cell infiltrates, vascular intimal thickening, and vessel narrowing. These vessels gradually lose their elasticity, and the media and adventitia become fibrotic [2]. Pathogenesis of disease suggests perivascular infiltrate and vasculitis, but is limited, as biopsies are not routinely done. In a few reported cases, histological findings include gliosis, leptomeningeal band-like sclerosis, thickened blood vessel walls, and intraparenchymal calcifications, all of which suggests a chronic inflammatory process [2].

In conclusion, typical neuroimaging features of linear scleroderma including atrophy, white matter lesions and calcifications typically involve the cerebral hemisphere ipsilateral to the skin

lesion. It is important to recognize that even in the absence of neurological symptoms, patients with linear scleroderma may present with neuroimaging features found on routine imaging.

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