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Case Report

Isolated Linear Scleroderma of the Face and Scalp: A Rare Variant

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Abstract: A kind of localised scleroderma known as "En coup de sabre" is characterised by band-like sclerotic lesions that usually affect the frontoparietal regions of the scalp. There are two variations of linear morphea in the head and neck that may be linked to neurologic symptoms known as En coup de Sabre and Parry-Romberg syndrome. On imaging, patients may have lesions in the cerebrum ipsilateral to scalp abnormality. Here, we discuss the neuroimaging of a 12 year-old boy who presented with seizures and had a right frontoparietal "en coup de sabre" scalp lesion.

Keywords: En coup de sabre (ECDS), Linear morphea, Neurologic symptoms, Neuroimaging, Localized scleroderma.

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INTRODUCTION

Localised scleroderma has a rare subtype called linear scleroderma "en coup de sabre" (ECDS). A rare condition known as localised scleroderma affects both adults and children [1]. Based on clinical characteristics, LS can be differentiated from systemic sclerosis, commonly known as systemic scleroderma. The absence of the following findings is a characteristic of LS: Internal organ involvement, nailfold capillary alterations, digital sclerosis, digital necrosis, and Raynaud syndrome [2]. Although epilepsy is the most prevalent neurologic symptom, there have been reports of other neurologic deficiencies such as mobility difficulties or behavioural abnormalities. Neurologic disorders and cerebral inflammation may be linked to both PRS and ECDS. Numerous neurological problems, most frequently headaches and seizures, have been documented. A widely used clinical classification system described by Peterson *et al*., subdivides LS into five subtypes: plaque morphea, generalized morphea, bullous morphea, linear scleroderma, and deep morphea. Furthermore, magnetic resonance imaging (MRI) is recommended for the identification of intracerebral alterations [1, 2]. Imaging abnormalities include brain atrophy, white matter lesions, intracerebral calcification, meningeal changes and skull atrophy which are mainly located on the same side as the skin lesions but can also occur on the opposite side [3]. No standard treatment regimen exists for localized craniofacial scleroderma. For isolated cutaneous involvement, treatments include topical corticosteroids, phototherapy, methotrexate and glucocorticoids [4]. The aim of this report is to provide additional imaging perspectives for this rare diagnosis.

CASE REPORT

A 12 year old boy was referred to Department of Radiodiagnosis after presenting with seizures. On physical examination, patient had a band like scalp lesion in the right frontal and parietal region. There is accompanying hair loss and right facial hemi-atrophy. No members of his family has similar lesions.

An MRI Brain with contrast was performed on GE signa pioneer 3T machine. At the time of diagnosis, imaging demonstrated focal soft tissue volume loss in pre-auricular and infra zygomatic region on right side, predominantly involving the subcutaneous fat with almost complete loss of subcutaneous fat. No definite enhancement made out in the region. There is no significant volume loss in underlying muscle.

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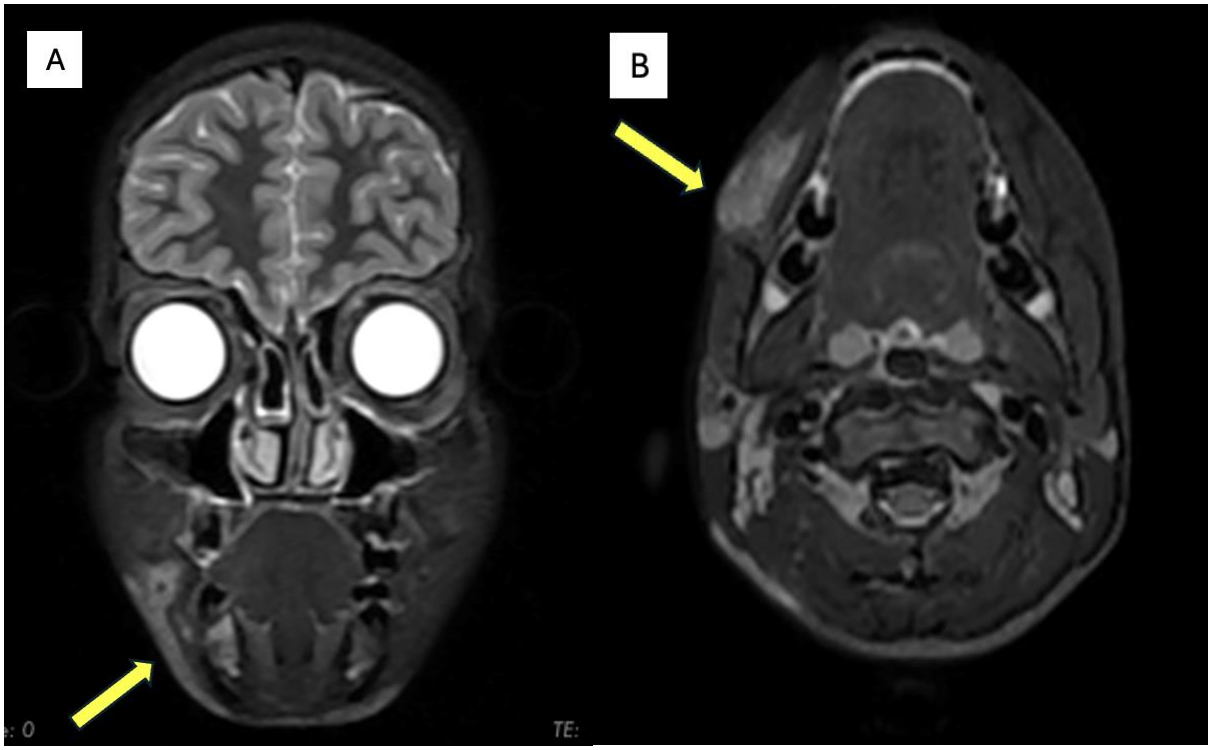

Figure (A) Coronal STIR image demonstrated altered signal intensity in the right mandibular and infra mandibular regions with loss of subcutaneous fat. Figure (B) Axial STIR image shows altered signal intensity in the right mandibular region with loss of subcutaneous fat

Focal altered signal changes (STIR hyperintense signal) noted in subcutaneous fat plane in the right pre-auricular and supra-zygomatic regions, measuring ~ 38 x 5 mm.

Similar altered signal changes seen in right mandibular and infra-mandibular regions, measuring ~ 36 x 10 mm (Figure A and B) This signal change also seen extending across the midline into the left infra-mandibular subcutaneous fat.

No obvious signal change in underlying muscles.

He brain of the patient showed no atrophy of brain parenchyma or abnormal signal intensity.

DISCUSSION

Localised scleroderma (LS), commonly known as morphea, is an inflammatory condition that causes atrophic and diffuse fibromatosis of the skin. Furthermore, LS may have an impact on musculoskeletal structures [1,2]. Localized scleroderma is differentiated from systemic sclerosis by the absence of sclerodactyly, Raynaud's phenomenon, capillaroscopic abnormalities, and organ involvement [1]. The mechanism by which the neurological system is involved in linear scleroderma is the subject of various theories. Endothelial cell death triggers fibroblast activation, which is followed by collagen contraction and cell cavity narrowing, both of which contribute to local ischaemia. Second, inflammatory lesions, particularly angio inflammatory lesions, have an impact on the nervous system [3]. Several authors have postulated that ECDS and PHA may be clinical variants of the same disease. 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 [3, 4]. Epilepsy and headache are the most common symptoms of linear scleroderma of the head and face involving the nervous system. The linear subtype of localized scleroderma often manifests in a en coup de sabre distribution, resembling fronto parietal laceration [3-5]. Facial atrophy occurs if the underlying muscle, cartilage, and bone are involved [1]. Skull and intracranial abnormalities can be seen on CT and MRI images, and imaging alterations can happen even in patients who are asymptomatic.The following imaging abnormalities are typically found on the same side as the skin lesions, but they can also occur on the other side: brain atrophy, white matter lesions, intra-cerebral calcification, meningeal alterations, and skull atrophy. In this instance, there was no brain parenchymal changes but there was ipsilateral subcutaneous fat loss and altered signal changes [3]. 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 anti-nuclear antibodies, antisingle-stranded DNA antibodies and rheumatoid factor [1].

CONCLUSION

Linear scleroderma, though a localized form of scleroderma, can have deep and functionally significant involvement, particularly when affecting the face or extremities. Early diagnosis, aided by clinical assessment and imaging, is essential to evaluate the extent of skin and soft tissue involvement. This case highlights the importance of multidisciplinary management involving dermatology, rheumatology, radiology, and physical therapy is key to optimizing patient outcomes.

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