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Case Report

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A CASE SERIES OF VARIED PRESENTATIONS OF A RARE ENTITY- LINEAR SCLERODERMA "EN COUP DE SABRE"

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ABSTRACT

Scleroderma is a complex autoimmune collagen disorder that can affect many organs simultaneously, as it occurs in systemic sclerosis, or only the skin, as it occurs in localized scleroderma. Linear scleroderma, a subtype of localised scleroderma (morphea) is termed as "en-coup de sabre" when it occurs on the scalp or forehead. Neurological involvement is rare. We report three cases of young patients in second to third decade of life. These patients had linear depression over the forehead and two of them had history of well controlled seizures.

KEYWORDS: Linear Scleroderma, MRI Brain, seizures.

INTRODUCTION

Linear scleroderma is a subtype of localised scleroderma (morphea). It is traditionally considered to be limited to skin, subcutaneous tissue, underlying bone, and, in craniofacial subtype nervous system involvement.[1] It is termed as "en-coup de sabre" when it occurs on the scalp or forehead. Neurological involvement is rare.Incidence is 0.4 to 2.7 per 100,000 people[1] with increased prevalence in the white population.[1] Mean age of onset is 13 years.[2] Females are predominantly affected.[3]

CASE SERIES

Case 1: 23 year old female patient with history of epilepsy since 9 years well controlled on medicinespresented with progressive depression on the left forehead (fig.1a). There was no history of trauma.

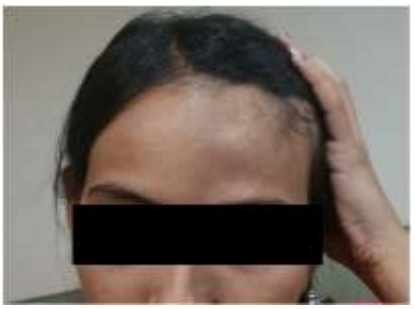

Figure 1(a): Showing linear depression on the left forehead.

CT brain with 3D reconstruction shows thinning of the subcutaneous tissue and underlying bone over left frontal region. (fig 1b and 1c).

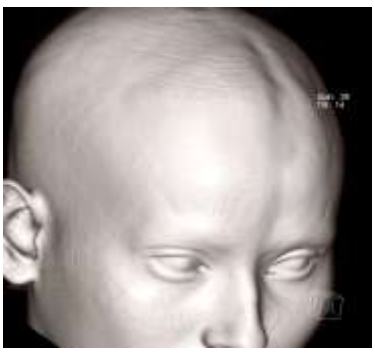

Figure 1(b).

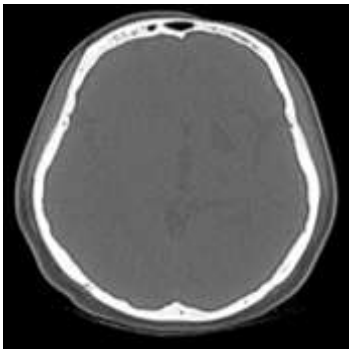

Figure 1(c).

The first MRI in 2009 showed multiple hyperintensities in left frontal subcortical and periventricular white matter on T2W (fig.1d) and FLAIR axial images (fig.1e).Thinning of the subcutaneous tissue over the left frontal region is seen on T1W axial image (fig. 1f).

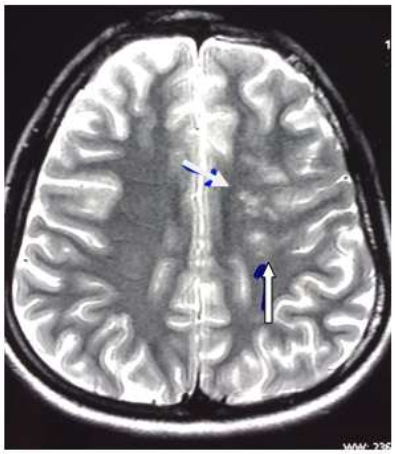

Figure 1(d).

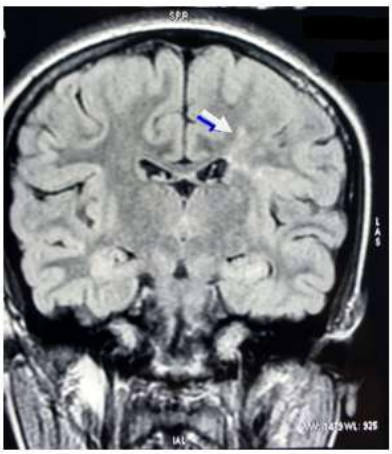

Figure 1(e).

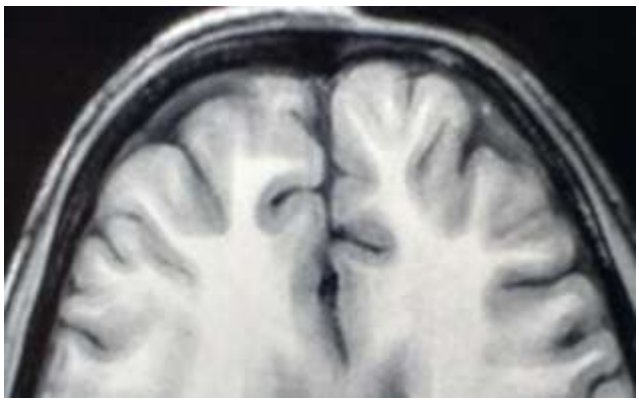

Figure 1(f).

A repeat MRI was performed in 2017 which showed progression of the hyperintensities in the white matter with new area of gliosis in the basal ganglia and ex-vacuo dilatation of the left lateral ventricle on FLAIR axial images. (fig 1g and 1h).Multiple foci of T2 shortening were seen in the subcortical white matter on left side on GRE images (fig 1i and 1j) . MRA reveals normal calibre of intracranial vessels. (fig 1k).

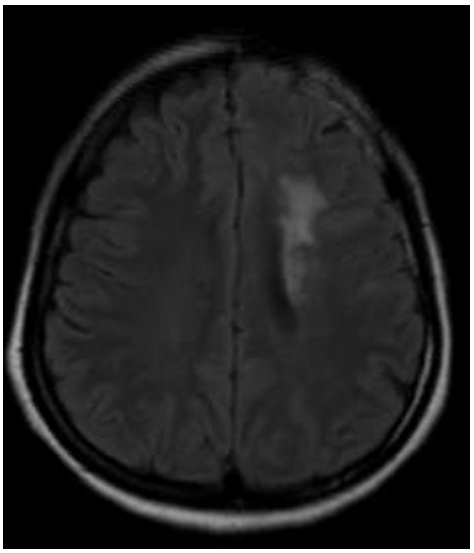

Figure 1(g).

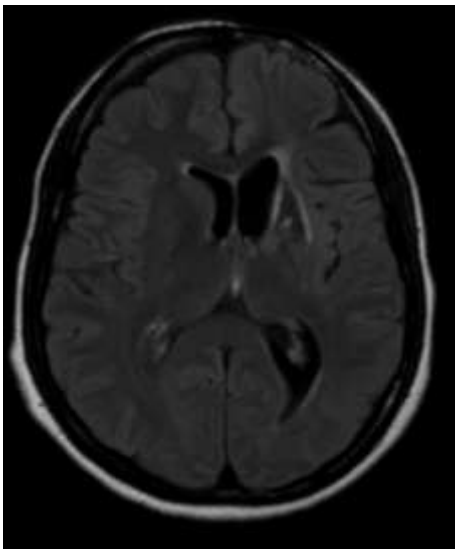

Figure 1(h).

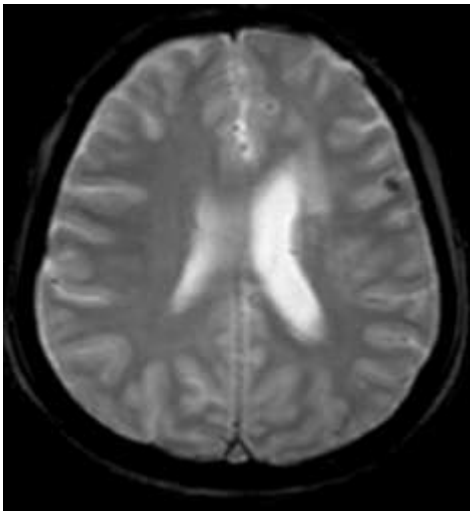

Figure 1(i).

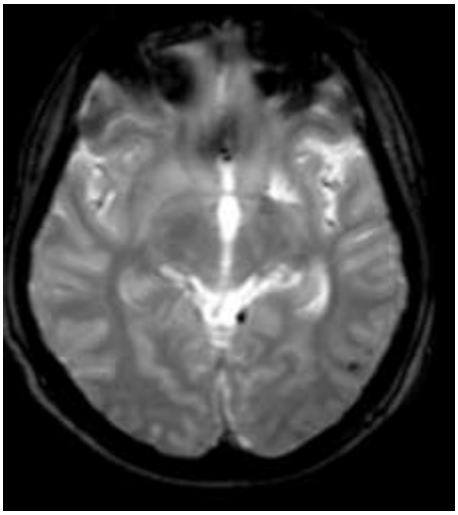

Figure 1(j).

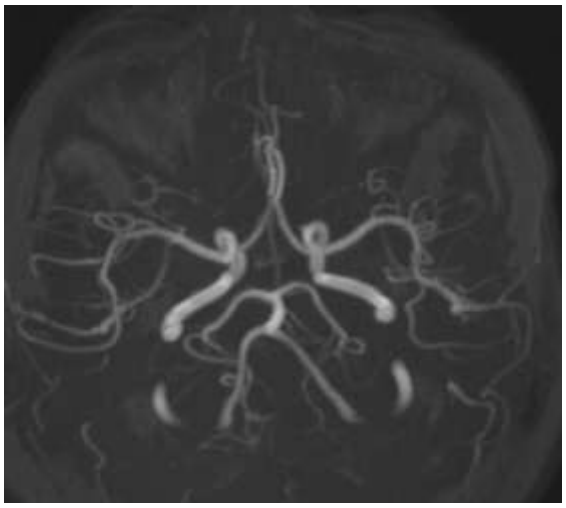

Figure 1(k).

Case 2: 18 year old male patient presented with an episode of seizure and linear defect over the left forehead without any history of trauma.

MRI performed showed thinning of the subcutaneous tissues and underlying bone over the left frontal scalp region on T1W axial and sagittal images. (fig 2a and 2b)White matter hyperintensities were seen in the left centrum semiovale and corona radiata on FLAIR axial images. (fig 2c and 2d).

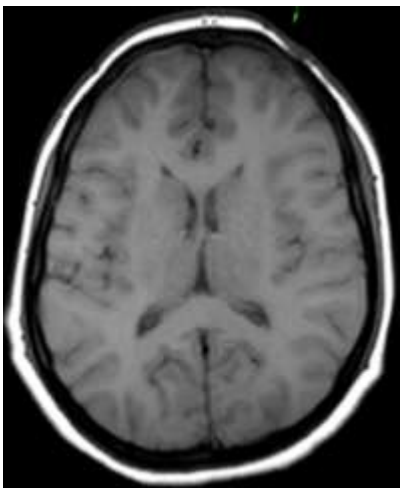

Figure 2(a).

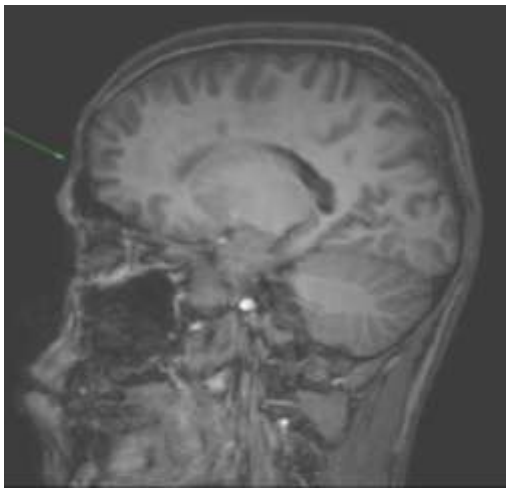

Figure 2(b).

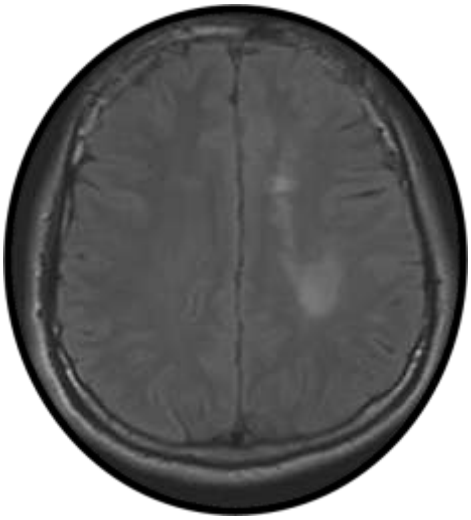

Figure 2(c).

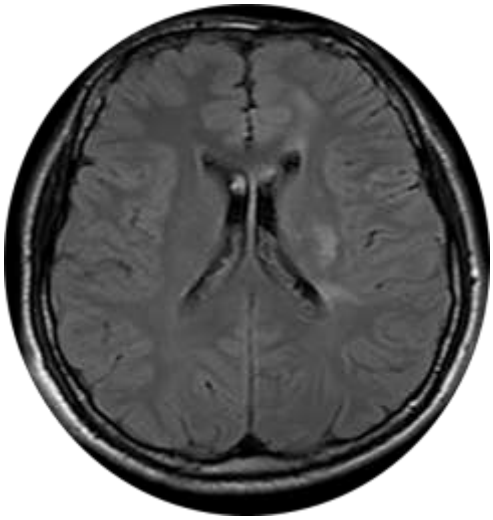

Figure 2(d)

Case 3: 16 year old female with linear area of depression over the scalp with no history of seizures or trauma.

MRI was performed which showed loss of subcutaneous tissue and bone on T1W sagittal image (fig.3a) with no white matter findings on FLAIR axial image (fig.3b).

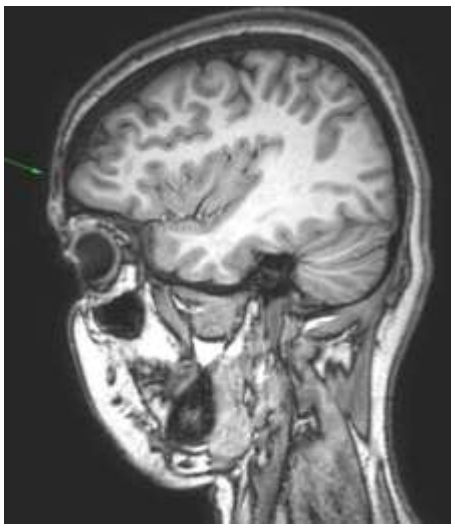

Figure 3(a).

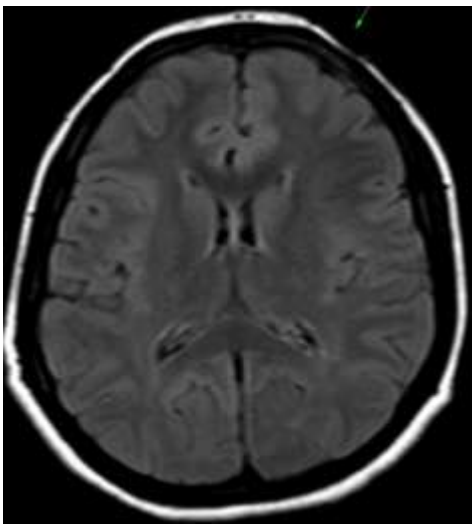

Figure 3(b).

DISCUSSION

Scleroderma is a rare disease of unknown etiology, characterized by thickening and hardening of skin resulting from increased collagen production. The term includes a variety of diseases, from localized scleroderma (LS) to systemic sclerosis. LS is traditionally considered to be limited to skin, subcutaneous tissue, underlying bone, and, in craniofacial subtype, nervous system involvement.[1] Linear scleroderma or en coup de sabre (LCsc) is a rare subset of LS. The typical presentation affects frontoparietal region, and the mean age of onset is around 13 years.[1]

Incidence is 0.4 to 2.7 per 100,000 people[1] with increased prevalence in the white population.[1] Females are predominantly affected.[3]

LScs has been associated with a variety of neurological abnormalities and typically is preceded by the development of cutaneous disease by months to years.[4] Neurological symptoms and signs include epilepsy,[4] headache, focal neurologic deficits, and movement disorders.[5,6]

Pathogenesis involves increased vascular permeability associated with mononuclear cell infiltration, leading to perivascular inflammatory cell infiltrates, vascular intimal thickening, and vessel narrowing.[4] Gradually, the vessels lose their elasticity; media and adventitia become fibrotic and more prone to small-artery occlusion.[4]

Computed tomography (CT) and magnetic resonance (MRI)studies have shown central nervous system abnormalities in LScs patients. Neurologic findings are

more frequently ispilateral to the skin lesions, but contralateral involvement has been described.[2,7] Outer diploe thinning, cerebral atrophy, white matter lesions, focal subcortical calcifications, and meningocortical alterations have been described.[2,8]

MRI usually exhibits T2 hyperintensities, mostly in subcortical white matter, but also in corpus callosum, deep grey nuclei, and brain stem.[2,9] Cerebral atrophy is generally subtle, characterized by blurring of the gray-white interface, cortical thickening and abnormal gyral pattern.[2,9] Atrophy is usually focal but widespread lesions involving an entire cerebral hemisphere have been described.[2,8] Infratentorial lesions and cerebellar hemiatrophy have been observed in patients presenting more severe neurological symptoms.[5]

Cerebral angiograms and magnetic resonance angiograms studies showed vascular involvement suggestive of vasculitis. Reports of cerebral aneurysms and other vascular malformations, as brain cavernomas , exist and could represent late sequelae of vasculitic process.[2,10,11]

At present no specific treatment is available .In reported cases, administration of methotrexate or mycophenolate mofetil and steroids appeared to have impact in controlling intractable seizures and stabilizing central nervous system damage.[2,12]

CONCLUSION

The investigations of choice are CT, to detect skull abnormalities, and MRI, to identify underlying brain lesions. Neuroimaging studies should be considered in all LScs patients at the time of the diagnosis.White matter changes and areas of T2 shortening ipsilateral to the subcutaneous fat loss and bone thinning suggest neurological involvement. Despite the patient being clinically asymptomatic, MRI may show progressive white matter changes. Hence, longitudinal studies should be done to look for progression.

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