Journal of Pakistan Association of Dermatologists. 2021;**31(4)**:731-734.

Case Report

Neurological manifestations of progressive hemifacial atrophy - A case report

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Abstract Progressive hemifacial atrophy is a rare, unilateral, atrophic craniofacial form of morphea involving the dermis, subcutaneous tissue, fat, muscle, and even bone. We report a case of a child having progressive hemifacial atrophy, presented with rapidly progressive neurological symptoms and associated radiological manifestations. In this case of hemifacial atrophy, the sensitivity of MRI in detecting intracranial lesions is demonstrated. Recommendations for imaging these patients are proposed.

Key words
Progressive hemifacial atrophy, neurological findings.

Introduction

Parry-Romberg syndrome, also known as progressive hemifacial atrophy (PHA), is a rare disorder characterized by unilateral facial atrophy affecting the skin, subcutaneous tissue, muscles, and sometimes extending to the osteocartilaginous structures.[1] The major features of this syndrome are atrophy of the soft tissues on one side of the face with hyperpigmentation of the overlying skin and neurologic findings.[2] Neurological complications are rare, they include Hemianopia, Facial palsy, Aphasia, Facial neuralgia, Hemiplegia, nerve palsies and behavioral changes.[3]

Case report

A 12 year old boy, known case of progressive hemifacial atrophy for 10 years, presented with complaint of sudden onset of right sided weakness and aphasia for 3 days. He was on different treatment modalities during past 10 years. From last 3 years he was taking oral Methotrexate 15mg/week but he was not compliant to medicine. Now he presented with sudden weakness of right side of body and aphasia. There was no history of fever, headache, neck pain, vomiting, seizures, blurring of vision. On Examination patient was irritable, and had GCS 12/15 (E4V2M6). Power in right upper and lower limbs was 3/5. Tone, muscle bulk and reflexes were normal. Positive Babinski sign on the right side. Examination of Left upper and lower limb was normal. Neck stiffness, Brudzinski's and Kernig's sign were negative. Other systemic examination was normal. Complete blood count and peripheral smear showed microcytosis and hypochromia. Other baseline investigations were normal. CT brain showed nodular and linear calcifications deposited over left frontal gyri along with hypoattenuated regional white matter including anterior limb of internal capsule. Marked thinning of left frontal bone in left para sagittal location.

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Figure 1, 2 Left facial atrophy with hyperpigmented, linear, atrophic, depressed scar extending from forehead upto chin.

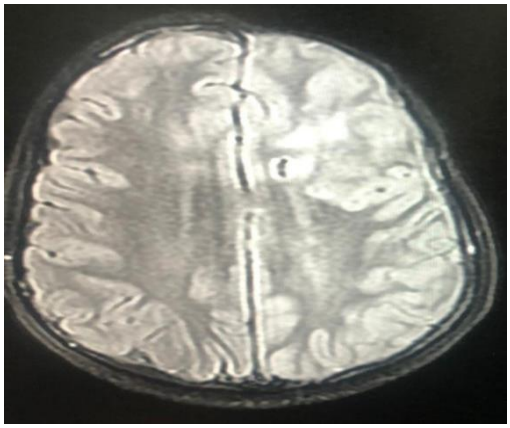

Figure 3 Irregular convulations, white matter signal abnormality and grey matter thickening in left frontal lobe.

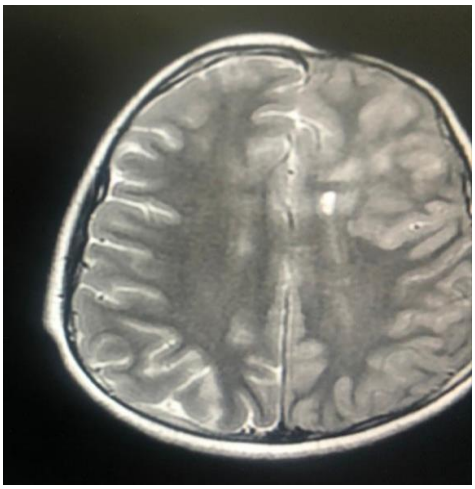

Figure 4 MRI axial T2 FLAIR: left frontal lobe hyperintense lesion and thinning of left frontal bone along with underlying gyri thickening.

MRI Brain showed abnormal grey matter thickening, irregular convolutions and white matter signal abnormality in left frontal lobe with mild volume loss as compared to right side. Old lacunar infarcts in medial temporal lobe, genu of corpus callosum and left centrum semiovale. There were micro hemorrhages in both cerebral hemispheres predominantly on left. Also there were foci of calcifications in left anterior frontal cortex and thickening of left tentorium cerebelli.

Patient was given intravenous dexamethasone 4mg daily. He showed marked improvement in power and speech after 3 days of treatment. Because of the rapid clinical improvement we did not undertake a brain biopsy. After 1 week of treatment , patient's GCS was 15/15, power of right upper and lower limb was 5/5. Patient was discharged on methotrexate 10mg/week and low dose oral steroid and was advised regular followup in dermatology and neurology outpatient department.

Discussion

PHA is a slowly progressing facial atrophy of subcutaneous fat and the wasting of associated skin, cartilage, and bone. This disorder includes an active progressive phase (2 to 10 years) followed by a burning out of the atrophic process with subsequent stability.[4] It effects the area supplied by one or multiple branches of the trigeminal nerve. Histopathology shows homogenized dermal sclerosis, fat atrophy, decrease in adnexal structures, and perivascular plasma cells and lymphocytes.[5]

Neurological manifestations are rare. They occur in only 4.4% of patients with linear morphea involving scalp.[6]

Epilepsy is the most common cerebral manifestation (60.5%). Headaches represent 44.2% of the neurological symptoms.[7] Other less frequent manifestations include trigeminal

neuralgia, cranial nerve palsies, limb weakness, aphasia, hallucinations, nystagmus and cognitive abnormalities.

In our patient, the disease remained localized for 10 years. MRI brain done 2 years back was normal. Now suddenly patient developed neurological symptoms. MRI showed abnormal grey matter thickening, irregular convolutions and white matter signal abnormality in left frontal lobe with mild volume loss as compared to the right side. There were Old lacunar infarcts in medial temporal lobe, genu of corpus callosum, left centrum semiovale and micro hemorrhages in both cerebral hemispheres predominantly on left. Also there were foci of calcifications in left anterior frontal cortex and thickening of left tentorium cerebelli.

Hence noninfectious inflammatory process causing chronic vasomotor disturbance and sympathetic nerve chain inflammation, is a major factor in the pathogenesis of this syndrome.[8]

Treatment of PHA can be challenging. Methotrexate (MTX) is the standard therapy. It is given in dose of 0.3-1 milligrams/ kilogram/ week (mg/kg/wk) with a maximum dose of 25 mg weekly in either an oral or injectable form. Oral prednisone is given over the first three months due to the fact that the methotrexate has a delayed effect on inflammation and fibrosis.[9]

Other immunosuppressant agents such as cyclophosphamide, cyclosporine, D-penicillamine, rituximab, and hydroxychloroquine are used when more aggressive therapy is required.[10]

Treatment may require the coordinated efforts of a team of Pediatricians or internists, surgeons (especially plastic surgeons), dentists, ophthalmologists, dermatologists, neurologists.

To halt progression of neurological symptoms, we started intravenous steroid and continued methotrexate. Patient showed marked improvement within 3 days. Patient was discharged after complete recovery of neurological symptoms and was advised to have regular follow up in Dermatology and Neurology outpatient department.

Conclusion

As progressive hemifacial atrophy is a rare, slowly progressive mutilating disease with involvement of brain parenchyma. Long-term follow-up of PRS patients is required, given the evidence that neurologic abnormalities may occasionally develop at any time over the course of the disease MRI sometimes reveals early abnormalities of the white matter even in patients without neurological symptoms and may be more sensitive than CT in the diagnostic evaluation of patients with progressive hemifacial atrophy. So serial MRI should be done to diagnose and treat brain involvement at early stage.

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