Case report

Parry Romberg syndrome: a case report

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Abstract:
Parry Romberg syndrome is a rare neurocutaneous disorder of unknown etiology characterized by facial hemiatrophy of fat, skin, connective tissue and MRI changes in the brain. Here we report a 30 years old woman who presented with facial hemiatrophy, headache and facial pain, hemi-masticatory spasm, atrophy of tongue, temporal hemianopia and hyperintense lesions on MRI.

Keywords: Parry Romberg syndrome; hemifacial atropy

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Introduction:

Parry Romberg syndrome (PRS, progressive facial hemiatrophy) is a neurocutaneous syndrome described first by PARRY[1] in1825, later by ROMBERG[2] in 1846. Age of onset of PRS is between 5-15 yrs. Prevalence is at least 1/7,00,000[3], with higher prevalence in females[4]. It is characterized by, atrophy of skin and subcutaneous structures such as fat, fascia, cartilage, bone, and /or muscles of one side of face.

Neurological manifestations are trigeminal neuralgia, migraine, seizures, with hyperintense lesions in grey and white matter of brain on MRI. Ocular manifestations are enophthalmos, ptosis, miosis, anhidrosis, and visual abnormalities. Oral manifestations include atrophy of tongue, dental abnormalities and hemi-masticatory spasm.

We present a case with features suggestive of PRS, with some atypical features.

Case report:

A 30yr old woman presented with progressive atrophy of right side of face for the past 8yrs. And she had recurrent episodic pain right side of head and face. Later she noticed paresthesiae, decreased sensation on right side of face and decreased hearing in right ear.

On examination, there is asymmetry of the right side of face, with atrophy of cheek and mandibular region. On nervous system examination, she has right eye temporal hemianopia, diminished corneal reflex and decreased sensation on right side of the face, sensory neural deafness in the right ear with lateralization

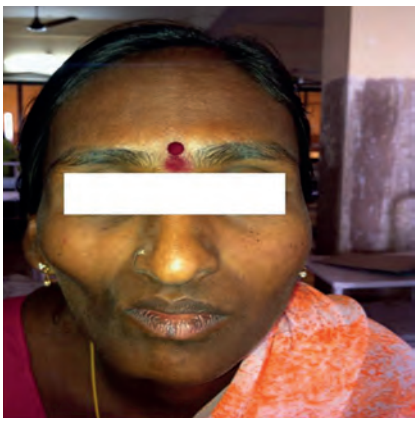

Figure 1 Facial Hemiatrophy: Front view

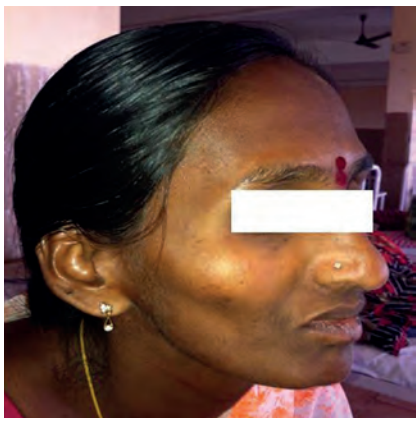

Figure2 Facial Hemiatrophy: Lateral view

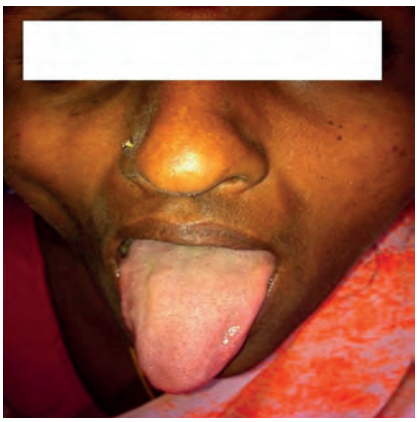

Figure 3: atrophy of tongue on right side

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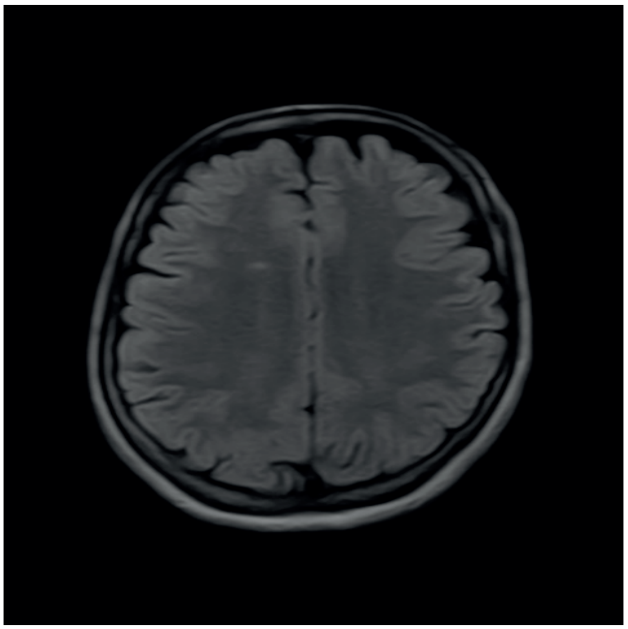

Figure 4 showing hyperintense lesions

to left ear on Weber`s test, atrophy of right half of the tongue, decreased sensation on right half of the body. No other neurological abnormality observed. Routine biochemical tests and hemogram were normal. Pure tone audiometry showed mixed hearing loss in right ear and MRI brain showed hyperintense lesions in the right frontal region on T2W images.

Discussion:

PRS is a rare disorder of unknown etiology. Average age of onset is around 10yrs. Common in females[4] in the ratio of 3:2. The prevalence is at least 1/700,000.[4] It is characterized by facial hemiatrophy of fat,skin, connective tissue, muscle, bone(100%), dental abnormalities(50%), migraine/facial pain(45%), jaw symptoms(including hemimasticatory spsm)(35%), atrophy of tongue(25%), vitiligo/ hyperpigmentation(20%), hemiatrophy of ipsilateral/ contralateral arm,trunk,leg(20%), epilepsy(10%), brain abnormalities on MRI(5%).[3] Contralateral hemianopia[5] is also described.

A global survey of 205 patients with PRS estimated the central nervous system involvement to be over 50%, with epilepsy in 11%.[6]

A case series on intracranial MRI appearances in ten patients with PRS,showed central nervous system involvement clinically and also on MRI in three patients, MRI abnormalities without clinical CNS involvement in two patients, no CNS involvement ,clinically or on MRI, in five patients. The MRI abnormalities consisted either of cerebral atrophy or white matter hyperintensities.[7]

Our patient is 30yr old woman, and presented with facial hemiatrophy, headache and facial pain, hemimasticatory spasm, atrophy of tongue, temporal hemianopia and hyperintense lesions on MRI, all of them representing a classical PRS.

But, atypical features in this case are, sensory loss on right side of face, right half of the body, hearing loss on right side. It is yet to be clarified whether they were coincident manifestations or were true association to PRS.

Conclusion:

PRS is an uncommon disorder, with unknown etiology, manifesting classically as atrophy of one side of face. And it should be considered in the differential diagnosis of all patients presenting with cranial nerve palsies especially, fifth and eighth, and hemisensory loss, if there is associated facial atrophy.

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