Commentary

It was in 1825 that Caleb Parry described the term progressive hemifacial atrophy, and the condition was better studied by Moritz Romberg in 1846.[1] Parry–Romberg syndrome (PRS) is characterized by progressive unilateral atrophy of the facial tissues leading to severe disfigurement with or without neurological complications. PRS may be associated with or without cutaneous involvement (linear scleroderma), en coup de sabre morphea, and neurological involvement with signs like seizures, trigeminal neuralgia, migraine headaches, ocular abnormalities, malalignment of jaw causing dental problems, and unilateral atrophy of tongue. In the internet survey done by Stone, 11% of patients self-reported a diagnosis of epilepsy along with PRS. PRS may be rarely associated with hemi-masticatory spasm (HMS).[2]

No randomized controlled trials have been conducted for PRS. The demographics and clinical findings of PRS are based on anecdotal case reports, small case series, and one internet survey. In the largest retrospective review of PRS patients by Tollefson *et al*.,[3] 68.5% were females, 57.4% patients presented with PRS at the time of presentation, and 27.8% had linear scleroderma or morphea initially followed by PRS. The mean age at onset was 13.6 years. 53.6% of the cases were associated with en coup de sabre and 13% had history of seizures. In the recent internet survey of 205 patients, the median age of onset was 10 years.[2] Bilateral facial involvement is very rare. Though PRS syndrome develops in the first or second decade of life, it can occur at any age.

Subcutaneous atrophy characteristically begins over the maxillary region, extending to chin, forehead, angle of mouth, and neck, and occasionally involves the ipsilateral half of the body. The overlying skin often becomes hyperpigmented, along with atrophy of the underlying bone and cartilage.[4] The pathogenesis of PRS is still unclear. It may be the result of arrested angiogenic process affecting the central nervous system during growth and development.

Neurological abnormalities like seizures, facial paresthesia, and abnormalities in computed tomography (CT) head or magnetic resonance imaging (MRI) have been associated with PRS. Association of PRS with HMS has been documented in several case reports.[5] HMS is a rare condition characterized by unilateral forceful contractions of one or more masticatory muscles. It is more commonly seen in females and not always associated with hemifacial atrophy. The proposed pathophysiology for the involuntary movement is peripheral irritation of the trigeminal nerve probably due to entrapment of the motor branches in the infratemporal fossa.[6] PRS with HMS onset and worsening during pregnancy has been documented.[7] The spasm usually improves after child birth. The exact mechanism of this occurrence in pregnancy is not known; probably it is related to hormonal changes during pregnancy.[7,8] Developmental delay with ipsilateral atrophy of the right hemisphere was documented in one neonate with progressive hemifacial atrophy.[9]

The relationship between PRS and en coup de sabre is debatable. Neurological abnormalities are also present in en coup de sabre. Some authors believe that there is significant overlap between the two entities, while some believe them to be two separate entities.[2,3] They represent differential spectrum of the same disease. Differential diagnoses include hemifacial microsomia and its variant Goldenhar syndrome, post-traumatic atrophy, and partial lipodystrophy (Barraquer–Simons syndrome). Hemifacial microsomia and Goldenhar syndrome are congenital and nonprogressive. In post-traumatic atrophy, history of trauma will be present. Lipodystrophy is usually bilateral and involves primarily adipose tissue.[4] HMS involvement in PRS should be differentiated from other causes like mechanical and inflammatory disorders of temporomandibular joint, tetanus and trismus, unilateral dystonia of jaw, hemifacial spasm, and facial myokymia.[10] In unilateral jaw dystonia, deviation of the jaw would persist for several days to weeks at a time before remitting, only to be followed by a further episode after a symptom-free interval.

Progressive stage with linear scleroderma can be treated with pulsed dexamethasone steroids with methotrexate and other immunosuppressants. When the disease is no longer progressive, surgical correction is done. Carbamazepine and botulinum toxin injection has been tried in HMS.[9] There are no diagnostic markers for disease progression or regression.

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References

1. Singh M, Bharija SC, Belhaj MS, Mangoush EA. Romberg's syndrome: A case report. Dermatologica 1985;170:145-6.
2. Stone J. Parry-Romberg syndrome: A global survey of 205 patients using the internet. Neurology 2003;61:674-6.
3. Tollefson MM, Witman PM. En coup de sabre morphea and Parry-Romberg syndrome: A retrospective review of 54 patients. J Am Acad Dermatol 2007;56:257-63.
4. Vykuntaraju KN, Sahana, Shivananda. Parry-Romberg syndrome. Indian Pediatr 2013;50:255.
5. Ebersbach G, Kabus C, Schelosky, L, Terstegge L, Poewe W. Hemimasticatory spasm in hemifacial atrophy: Diagnostic and therapeutic aspects in two patients. Mov Disord 1995;10:504-7.
6. Cruccu G, Inghilleri M, Berardelli A, Pauletti G, Casali C, Coratti P, *et al*. Pathophysiology of hemimasticatory spasm. J Neurol Neurosurg Psychiatry 1994;57:43-50.
7. Panda AK, Gopinath G, Singh S. Parry Romberg syndrome with hemimasticatory spasm in pregnancy; a dystonia mimic. J Neurosci Rural Pract 2014;5:184-6.
8. Cersósimo MG, Bertoti A, Roca CU, Micheli F. Botulinum toxin in a case of hemimasticatory spasm with severe worsening during pregnancy. Clin Neuropharmacol 2004;27:6-8.
9. Chang SE, Huh J, Choi JH, Sung KJ, Moon KC, Koh JK. Parry-Romberg syndrome with ipsilateral cerebral atrophy of neonatal onset. Pediatr Dermatol 1999;16:487-8.
10. Thompson PD, Obeso JA, Delgado G, Gallego J, Marsden CD. Focal dystonia of the jaw and the differential diagnosis of unilateral jaw and masticatory spasm. J Neurol Neurosurg Psychiatry 1986;49:651-6.

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