Parry-Romberg Syndrome (Progressive Hemifacial Atrophy) with Spasmodic Dysphonia - A Rare Association


Abstract

Parry-Romberg syndrome is a rare clinical entity characterised by progressive hemifacial atrophy with appearance of ‘saber’. Various neurological and otorhinolaryngological disorders are associated with this syndrome. The association of Parry-Romberg syndrome with Spasmodic dysphonia has rarely been reported. A 37 year old female presented with progressive atrophy of tissues of left side of face for 10 years and change in voice for 1 year. On examination, wasting and atrophy of tissues including tongue was noted on left side of the face. ENT examination revealed adductor spasmodic dysphonia. We report the rare association of Parry-Romberg syndrome with spasmodic dysphonia.

Introduction

Parry-Romberg syndrome is characterised by slowly progressive atrophy of subcutaneous tissue. Bones, muscles, eyes and brain may be affected by atrophy of unknown origin. Various neurological and otorhinolaryngological disorders are associated with this syndrome. As there are many otorhinolaryngological disorders are associated with Parry Romberg syndrome, spasmodic dysphonia can be a part of it. We present the case of Parry-Romberg syndrome with spasmodic dysphonia which is a rare association.

Case Report

A 37 year old female presented with slowly progressive wasting of the skin and soft tissues of left side of face for 10 years. Initially started in tissues above the upper jaw between the nose and the nasolabial fold, then progressed to the angle of mouth, lower jaw and the neck. She also gave history of difficulty in speaking with change in voice for the past 1 year. There was no history of seizures or facial pain.

On examination, she showed wasting and atrophy of the skin and soft tissues involving the left side of face(left cheek, nose, upper and lower lip and jaw) (Figure 1). There is also atrophy of tongue on the left side (Figure 2). In addition, there is hyperpigmented skin over left side of face involving left cheek, nose, upper and lower lip and jaw extending upto neck (Figure 3). She has difficulty in speaking in the form of difficulty to start words, stuttering of speech sounds and strained voice sounds suggestive of adductor spasmodic dysphonia. ENT opinion obtained, which revealed adductor spasmodic dysphonia with normal vocal cords. Based on the above findings Parry-Romberg syndrome(progressive hemifacial atrophy) with spasmodic dysphonia was made. Her EEG and MRI brain were normal. Panoramic radiography revealed no bony involvement of maxilla and mandible.

In view of spasmodic dysphonia, speech therapy and vocal cord relaxation therapy was planned. Failing which patient was counselled for botulinium toxin therapy. Patient underwent speech therapy and vocal cord relaxation therapy and found her vocalisation improved satisfactorily. On followup for a year, there is no further progression of facial atrophy.
Discussion

Parry-Romberg syndrome is a rare incurable craniofacial disorder characterised by slowly progressive atrophy of soft tissues of half of the face (Hemifacial atrophy). It was described by Parry (1825) and Romberg (1846). It occurs as either congenital failure of development or as a progressive atrophy of the skin, subcutaneous tissue and muscles of one half of face sometimes with trophic changes in the connective tissue, cartilage and bones. Loss of tongue muscles occurs in some patients. Accompanying changes include trophic changes in the hair with loss of pigmentation. The atrophy may cease abruptly or progress slowly and then become stable. The onset of disease is usually during 1st or early 2nd decade of life but occasionally begin during middle or later years, of life.

Different mechanisms have been discussed, immunologic disorders, sympathetic dysfunction or infectious origin in the pathogenesis of Parry-Romberg syndrome. Recently blink reflex and trigeminal evoked potential studies in these cases have indicated abnormalities in brain stem as well. Immunological evidence has favoured the possibility of involvement of noradrenergic system in the brain stem. Hyperactivity of the brain stem sympathetic centres, possibly caused by an autoimmune process may be the primary cause for cutaneous and subcutaneous atrophy in Parry-Romberg syndrome. The disease may be a neural crest defect.

Neurological features include migraine/trigeminal neuralgia (45%), epilepsy (10%) sometimes associated with ipsilateral brain changes in MRI (5%). There may be associated otorhinolaryngological disorders in the form of unilateral hearing loss, frontal sinus deformity and parotid gland atrophy in Parry Romberg disease. As there are many otorhinolaryngological disorders associated with Parry Romberg syndrome, spasmodic dysphonia can be a part of it.

In Parry-Romberg syndrome, there is paramedian atrophy without induration of the skin overlying the scalp with atrophy extending down the side of face often involving the tongue. In linear scleroderma, there is induration of the skin in the region of scalp which usually does not extend below the forehead and it is not associated with neurological abnormalities. In a full-fledged case of Parry Romberg syndrome, there is significant deformity, with one entire side of the face smaller than the other. This is in sharp contrast to typical linear scleroderma, where the abnormality is confined to the forehead. The difference between linear scleroderma and Parry Romberg syndrome is best exemplified by involvement of the tongue in Parry Romberg syndrome. Not only is one side of the face smaller, but also one side of the tongue is smaller. Thus, Parry Romberg syndrome is clearly not a condition of the skin alone. Linear scleroderma should not extend below the eyelid.

Histology of both Parry-Romberg syndrome and linear scleroderma is similar. However in case of linear scleroderma there is massive lymphocytic infiltration around the vessels of surface and deep plexus of skin. In Parry-Romberg syndrome, there is fibrosis, collagen fibre thickening, skin oedema and vessels atrophy without inflammatory infiltration.
Differential diagnoses include hemifacial microsomia (first and second branchial arch syndrome) and its variants, such as Goldenhar syndrome, but these are congenital and essentially non-progressive conditions. Post-traumatic atrophy and partial lipodystrophy (Barraquer-Simon Syndrome) are also included in the differential diagnosis. However, partial lipodystrophy is usually bilateral and involves primarily the adipose tissue. 7

Management of Parry-Romberg syndrome consists of reconstructive surgery, symptomatic treatment with psychological issues.

This case highlights the association of Parry-Romberg syndrome with spasmodic dysphonia which is rare.

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References