Facial Hemiatrophy: Review of Literature and a Case Report

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ABSTRACT

A case report of hemifacial atrophy is presented in this paper. It is also known as Parry-Romberg syndrome, is an uncommon degenerative and poorly understood condition. It is characterized by progressive atrophy of the skin, subcutaneous tissue, muscle, cartilage or bone; the condition can leave a marked deformity on one side of the face. The incidence and the cause of this alteration is debatable. The most common complications that appear in association to this health disorder are: trigeminal neuritis, facial paresthesia, severe headache and epilepsy. Now, plastic surgery with graft of autogenous fat can be performed, after stabilization of the disease, to correct the deformity. Orthodontic treatment can help in the correction of any associated malformation. This presentation gives a review of concern literature about the etiology, physiopathology, differential diagnosis and treatment of hemifacial atrophy.

Keywords: Hemifacial atrophy, Parry-Romberg syndrome.

INTRODUCTION

The first published reports on progressive hemifacial atrophy were attributed to Parry in 1825 and Romberg in 1846. It is characterized by progressive atrophy of the skin, subcutaneous tissue, muscle, cartilage or bone; the condition can leave a marked deformity on one side of the face. Hemifacial atrophy is also known as Parry-Romberg syndrome. It is characterized by a slow and progressive atrophy affecting one side of the face.

In 1927, Kraus and Perkins reviewed the literature on hemiatrophy and collected 18 cases, of which only five were not associated with motor disorders, such as hemiplegia and athetosis. They classified hemiatrophy into congenital, familial and acquired types. The hemiatrophy may be partial or complete. The sex distribution is equal. Hemiatrophy more commonly affects the left side of the body, as opposed to hemihypertrophy, which is commoner on the right side. Later, Landauer (1939) was able to collect 22 cases of partial or complete hemiatrophy, mostly in America. Mental defect and cutaneous abnormalities are common in cases of hemihypertrophy, but are rarely associated with hemiatrophy.

Hemiatrophy of the face is an uncommon deformity of unknown etiology. Two main types have been described; a congenital facial hemiatrophy and a progressive facial hemiatrophy (Walsh, 1949). The progressive facial hemiatrophy commences usually in the second decade and is seen more commonly in females than in males. The skin becomes thinned because of atrophy of the papillary layer and disappearance of the subcutaneous fat. Subsequently, the underlying cartilages, muscles and bones are affected. The maxillary, molar and palatine bones on the affected side are usually smaller and flatter. The mandible and the mastoid processes may be severely affected. The involvement of external ear is quite frequent. Purves-Stewart reported that the tongue, when protruded, comes out straight unlike that of a patient with atrophy from hypoglossal palsy. Duke-Elders stated that the patient may get speech difficulties as the tongue on the affected side may be underdeveloped. Atrophy of the tongue on the same side has also been reported (SHARP). The hair on the affected side may fall out or may become white.

We are reporting a case of a 24-year-old boy that has come for esthetic correction of asymmetry of face at the OPD, Oral Surgery, Peoples College of Dental Sciences, Bhopal. Detailed history and examination were done. Patient was asymptomatic at birth. He first noticed the deformity on the left side of face at the age of 5 to 6 years of age. The deformity increased till 14 to 15 years of age and ceased since then. Pain and paresthesia were not associated, also, there was no history of trauma or...
infections were recorded. No history of epilepsy or neuralgia given.

• Face—asymmetrical
• Lips—competent
• TMJ—movements bilaterally synchronous. No clicking/ deviation on opening/closing
• Skin color—no abnormality of pigmentation/vitiligo
• Ocular—no enophthalmos
• Patient was unable to blow air/whistle
• No trigger points for neuralgic pain
• No evidence of paresthesia
• Masseter—bulk reduced on left side
• Skin on cheek cannot be pinched
• Absence of subcutaneous fat/buccinator muscle noticed
• Teeth present 1 to 7 in all four quadrants
• No asymmetry of arch form noted.

OPG finding (Fig. 2) shows mandibular bone not symmetrical and hypoplasia of mandible is on left side. Angle of left side mandible is not prominent.

DISCUSSION

Authors Lanje et al in 2006 reported similar case of an 18-year-old girl. She had given similar history and we diagnose it as a case of hemifacial atrophy. Differential diagnosis for this we can consider posttrauma fat necrosis, Parry-Romberg syndrome, hemifacial microsomia, partial lipodystrophy, Goldenhar’s syndrome. But due to absence of trauma history posttrauma fat necrosis is not considered. Absence of contralateral Jacksonian epilepsy, trigeminal neuralgia, neuromuscular hypoplasia eludes the chances of classic Parry-Romberg syndrome. The deformity is not congenital.

OPG shows mandibular bone deformity but the micrognathia is not absolute or prominent feature. On intraoral examination, the teeth and tongue are normal. We excludes Goldenhar’s syndrome as our diagnosis as it shows absence of Oculoauculovertebral dysplasia. Wartenburg reported that cerebral disturbance may lead to increased and unregulated sympathetic nervous activity and produce localized atrophy. According to Poswillo (1973), facial deformity is result of disruption of stapedial artery. Robinson (1987) supported the theory of Poswillo. As previously reported, facial hemiatrophy is associated with ipsilateral salivary glands and hemiatrophy of tongue occurs. Extraoral manifestations show unilateral involvement of ear, larynx, esophagus, diaphragm, kidney and brain. Neurological deficits seen in 15% cases. Ocular deficits (enophthalmos) seen in 10 to 40% of the cases. Rarely, entire half of the body is seen to be involved with pigmentation disorder vitiligo, pigmented facial nevi, contralateral Jacksonian epilepsy, contralateral trigeminal neuralgia and ocular complication. As reports show facial atrophy shows more predilection for females (F:M—3:2). Slight predilection for left side occurs in first or second decade of life which is in accordance with our case. The deformity progresses over 2 to 10 years and atrophy appears to follow distribution of divisions of trigeminal nerve. Incomplete root formation, delayed tooth eruption, difficulty in mastication and hemiatrophy of lips and tongue are unusual findings. Nowadays, treatment modalities have been immensely improved. Correction of the soft and hard tissue defects can be done by using autogenous graft as well as alloplastic grafts.

REFERENCES