

Facial Hemiatrophy

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Facial hemiatrophy, also called Parry-Romberg syndrome, is a disease of unknown etiology characterized by slowly progressive atrophy of one half of the face with or without other associated features. Knowledge of this peculiar entity is essential for making a correct diagnosis because in its early stages the disease may mimic scleroderma or lipodystrophy. To the best of our knowledge, this is the first case report from India.

Case Report

A 9-year-old girl was born normally of a non-consanguineous marriage and was normal till 6 years of age. She then developed hypopigmented macules over trunk which started spreading progressively. She was given some Ayurvedic treatment after which the lesions became static but did not get repigmented. A year later she was noticed to have progressive wasting of the right side of the face, especially the jaw, with darkening of overlying skin. Within the next 6 months she developed gradual weakness and wasting of right arm leading to inability to straighten the elbow. She had no joint pain, dysphagia, fever, weight loss,

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or sensory symptoms. She had a normal appetite and was attending school regularly.

Examination revealed a thinly built child with marked wasting of right half of the body (*Fig. 1*). The right half of face was significantly smaller than the left (*Fig. 2*). The difference was most obvious when the right mandible was compared to the other side. The skin was puckered over the right mandible but was not tethered to the underlying tissue. There was a hypopigmented macule over the right temple, and hyperpigmented macules over the right temple, and the right half of the chin. The teeth, tongue, ears, and eyes were normal.

The right half of trunk and abdomen appeared wasted with the right scapula

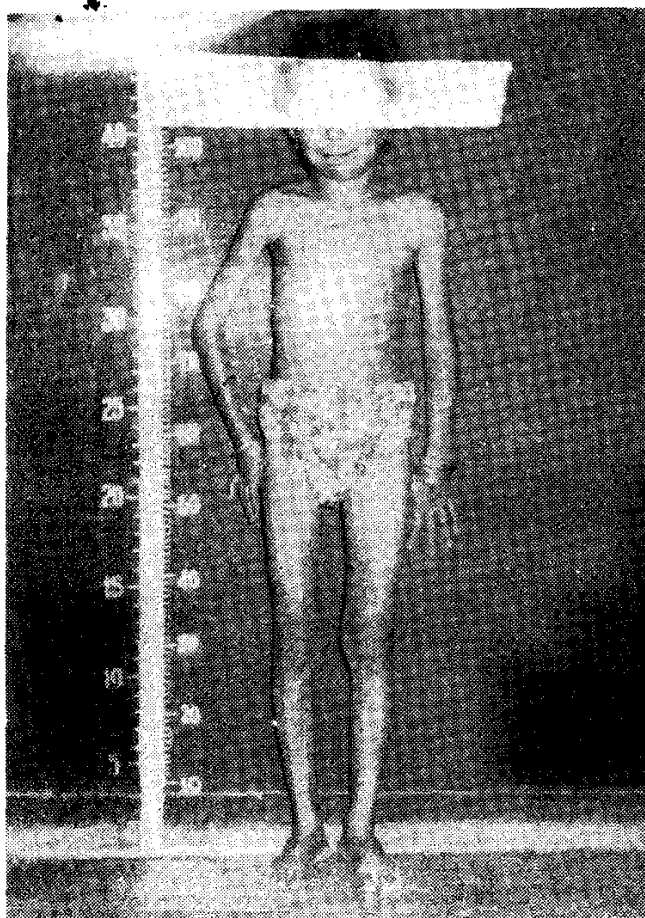


Fig. 1. Showing hemiatrophy of right side of body with deformity of right elbow.



Fig. 2. Showing atrophy of right side of face including jaw and increased pigmentation of skin on the right of jaw.

being smaller than the left. Here too there was puckering and wrinkling of the skin indicating loss of underlying tissue. The left trunk and abdomen showed hypopigmented scattered macules 1-5 cm in diameter in a segmental distribution. The right arm was smaller than left, with marked wasting and flexion contractures at the elbow. The right leg had muscle wasting and the sole of the foot was smaller than the left.

Neurological examination revealed that there was no sensory, motor or autonomic involvement on the right half of the body except weakness of right arm due to wasting. The optic fundi were normal.

Antinuclear factor was strongly positive indicating an underlying autoimmune process. Renal functions and urine examination were normal. CT scan of the head was normal and did not reveal any asymmetry. Ultrasonography showed that

the internal organs like ovaries and kidneys were of equal size bilaterally.

Discussion

Facial hemiatrophy was first described by Parry in 1825 and Romberg in 1846(1). The disease is characterized by progressive wasting of one side of the face. It may involve one side of the whole body or may even be bilateral. The wasting usually starts at the inner eye, forehead and angle of mouth, a few centimeters away from the midline. The affected side looks shrunken, old and wrinkled with premature greying and loss of hair. The affected tissues include skin, subcutaneous fat, connective tissue, bone, muscles and cartilage. Associated features include nevi, vitiligo and hyperpigmented areas on the affected side(2).

Autonomic involvement in the form of Horner's syndrome, decreased or increased sweating, and Argyll-Robertson Pupil have been reported. Homolateral hemicranial migraine, hemiplegia, hemianesthesia, hemi-anopia or epilepsy may be occasionally seen(1).

The disease is commoner in females than in males. It is usually sporadic although a familial case has been reported in a 7-year-old girl(3). It has to be differentiated from scleroderma. The unilateral distribution is, however, characteristic. Lipodystrophy may at times be considered, but bony involvement rules it out.

The diagnosis is based on the clinical picture. Investigations are non-specific. The affected tissues are atrophic with little evidence of inflammation. Muscle atrophy is predominantly due to loss of fat and connective tissue. The cerebral hemisphere on the affected side may be atrophic. Atrophy of homolateral internal organs including

kidneys, adrenals and ovaries, has been described(3).

A number of theories have been put forward to explain the etiology of this puzzling disease. The Trigeminal theory postulates that vascular insult to the trigeminal ganglion perhaps due to trauma to the superior cervical ganglion triggers off the wasting process leading to hemiatrophy(3). Casserier considers the disease to be a variant of scleroderma(3). Another plausible explanation is the Cerebral Trophoneurosis theory that there is a disturbance of higher centres causing unregulated activity of the cervical sympathetic system as a 'release' phenomenon. This has been supported by reports of pathological evidence of degeneration of nuclei in the diencephalic periaqueductal gray matter and lateral wall of the third ventricle(3).

The prognosis for life is good in this slowly progressive condition. The disease may smoulder for years to decades before spontaneous remission occurs. There is no definitive treatment except for plastic surgery to ameliorate the severe cosmetic disfigurement(4). This is usually undertaken after the disease has run its course, as determined by close clinical observation and monitoring of tissue breakdown enzymes(4). Mild to moderate cases need only dermal grafts. In severe deformity, transfer of de-epithelialized skin flaps may be undertaken. If the orbital, maxillary or mandibular bone are hypoplastic, bone grafting with or without osteotomy is carried out(5).

REFERENCES

1. Brain R. Disease of the Nervous System, 8th edn. London, Oxford University Press, 1977, pp 712-713.
2. Archambault L, Fromm NK. Progressive

2. facial hemiatrophy. Arch Neurol Psychiatr 1932, 27: 529-581.
3. Wartenberg R. Progressive facial hemiatrophy. Arch Neurol Psychiatr 1945, 54: 75-96.
4. Wallace JG, Schnuder WJ, Brown KG, *et al.* Reconstruction of hemifacial atrophy with a free flap of omentum. Br J Plast Surg 1979, 32: 15-18.
5. Jackson IT. Plastic surgery of face and neck. In: Practice of Pediatrics. Ed Kelly VC. Philadelphia, Harper and Row, 1985, pp 11-12.

Values for Foot Length in Newborns

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Abnormalities of foot length are important features of some syndromes that are recognizable at birth(1). Merlob *et al.* have set forth standards for foot length for caucasian newborn infants of different gestational ages(1). However, there are no such studies reported from India. Many babies with malformation syndromes are born prematurely and therefore, there is a need to know the range of values for foot length at different gestational ages. Another use of foot length measurement may be in the assessment of fetal growth(2). Because of the potential diagnostic implication of foot length measurement the present study was undertaken to know the range of values for the foot length in infants from 26-42 weeks of gestation.

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