FACIAL HEMIATROPHY OF ROMBERG AND PARRY - A CASE REPORT

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A case of facial hemiatrophy of Romberg and Parry is reported. The patient had all the changes of the disease with an interesting change in E E G.

Key Words : Facial hemiatrophy of Romberg and Parry, E E G changes

Introduction

First described by Parry in 1825 and Romberg in 1847 and named after them by Eulemberg in 1871, facial hemiatrophy is a disorder of subcutaneous tissue, muscles and bones.[1] A relationship to morphoea (localised scleroderma) has been postulated[2] but this could not explain the occasional involvement of other parts of the body on the affected side or the ipsilateral atrophy of the cerebral hemisphere.[3] There is no evidence suggestive of its genetic inheritance, but some cases showed that it may be hereditary.[4]

Case Report

A 25-years-old male patient presented with deformity and asymmetry of the right side of the face for the last 2 years. It started with the darkening of the right forehead which spread gradually to involve the right side of the face and a portion of the upper neck. He lost hairs on the right temple, right upper eyelid. Sweating was absent on the affected side for the same duration. There was no power deficit of the facial muscles of the right half. He gave no history suggestive of hereditary affection. There was no history of epilepsy or any other neurological deficit.

The skin of the affected side showed sharply margined, hyperpigmented, shiny nature. The symmetry of the face was lost, nose and chin were deformed, right eye was protruding from the orbit (Fig. 1). Teeth on the

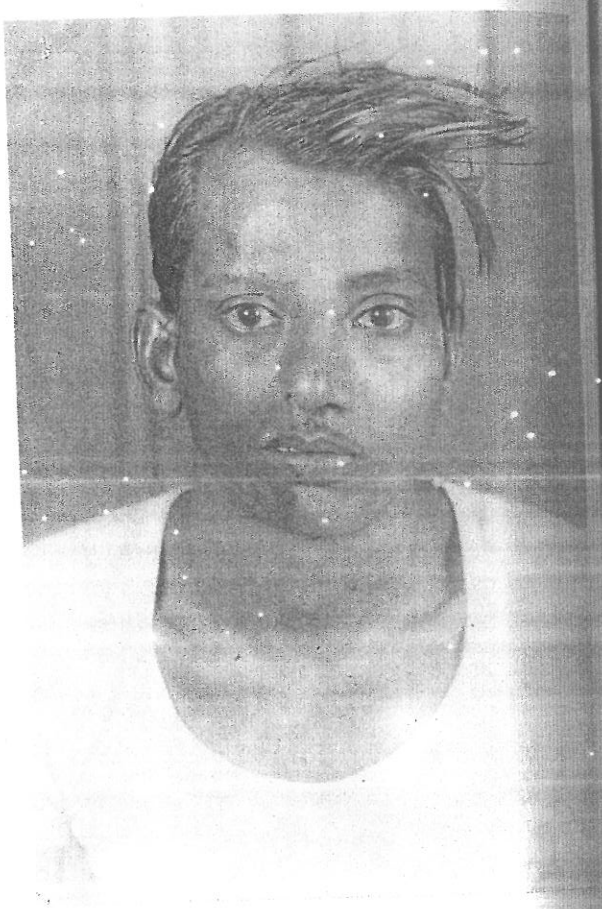

Fig. 1. Deformed facial appearance in facial hemiatrophy.

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right side were maloccluded. Right palate was also deformed but tongue was normal in appearance. Sensation on both sides of the face was normal.

Investigations, which included complete haemogram, liver function test, urine examination and stool examination were normal. Roentgenogram of skull,spine and barium swallow of the oesophagus showed no abnormality but X-ray (PNS) showed hypoplastic frontal sinus on the right side, smaller maxillary sinus on the right side (2.1 cms) compared to the left side (2.6 cms); increased radiolucency on the right side as compared to the left. E C G was normal; E E G showed generalised slow wave activity on the right cerebral hemisphere. C T scan was normal. Electromyographic study using concentric needle electrode was done to see spontaneous and voluntary activities of the frontalis and orbicularis oris muscles. This showed partial activity in the said muscles.

Histopathological examination of the section taken from the skin of the forehead showed only increased number of melanocytes in the basal layer, but no other change.

Comments

Facial hemiatrophy is more common in adolescent girls,[1] but in our case the patient is a male in his mid twenties. The hair may be lost in the frontoparietal region, but is often normal. In this case hair on the right side of face was lost. The disease usually progresses to the whole face, but occasionally upto the neck, as in this case and even upto the ipsilateral breast.[3] Steif (1953) described vasodilatation in the ipsilateral hemisphere. In a recent report of 3 cases, Asher and Berg (1982) found changes in the C T scan suggestive of vascular malformation in the ipsilateral hemisphere in 1, and ill defined abnormality in the opposite hemisphere in the 2, and normal C T scan in the 3. The cerebral hemisphere of the affected side may be atrophic.[3] Although in our case C T scan is normal but E E G shows features which may be suggestive of cerebral atrophy on the affected side, which is an interesting feature in the present case.

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