RESEARCH PAPER | Medical Science | Volume : 5 | Issue : 7 | July 2015 | ISSN - 2249-555X

A Case of Parry Romberg Syndrome

KEYWORDS: Parry Romberg syndrome, en coup de sabre, morphea

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ABSTRACT *Parry Romberg syndrome is a rare acquired poorly understood neurocutaneous syndrome of unknown etiology characterized by slow progressive atrophic changes commonly affecting one half of the face. The exact incidence and etiology remains unclear. Apart from the multifactorial etiology proposed, the possible primary cause is mainly attributed to the cerebral disturbance of the fat metabolism. The syndrome overlaps with "en coup de sabre"and morphea, with an ill defined relationship existing between the two. We describe a girl with localized cicatricial alopecia over frontal region of scalp with coup de sabre lesion. Biopsy from affected site is suggestive of morphea. These findings support the diagnosis of Parry Romberg syndrome*

CASE REPORT

13 year old girl presented with complaints of hyperpigmented patch over the right side of the forehead since birth which was gradually progressing in size without any associated symptoms. Over past five years mother noticed a depression under the skin which was also getting worse. There was history of focal alopecia since birth. There was no history of pain, trauma, unilateral headache, seizures, drooping of eyelid, redness of conjunctiva. She had achieved age appropriate milestones going to a normal school studying in eighth standard. Her birth history was normal and immunised for age without any similar illness in the family. Her height was 128 cms and she weighed 18.5 kg. On examination she had a hyperpigmented patch with atrophied skin on the right side of forehead with bony depression under it. With the above features , we had the differential diagnosis- Aplasia cutis, Goltz syndrome and Parry Romberg syndrome, dermatologist opinion was sought who described the lesion as 'En coup de sabre lesion' (Holland and Nocton) Circumscribed patch of scleroderma (Kaplan MJ) on right side of forehead & lateral aspect of right cheek with - Cicatricial alopecia on right side of scalp.

Skin biopsy was done which revealed skin with unremarkable epidermis with underlying thin dermis showing hyalinisation of collagen in the superficial dermis and skin adnexal structures with underlying subcutaneous tissue showing mature adipose tissue with interspersed collagen fibres (Kaplan MJ). Features suggestive of morphea (Kaplan MJ) which were consistent with Parry Romberg syndrome.

CT BRAIN: Mild thinning of outer and inner table and loss of diploic space of right frontal bone with adjacent soft tissue scalp thinning noted. No evidence of bony erosions/defect. No significant intracranial abnormality.

INTRODUCTION

Parry-Romberg syndrome is a rare disorder characterized by slowly progressive atrophy of the skin and soft tissues of half of the face (hemifacial atrophy), usually the left side. It occurs more commonly in females than in males. Initially there is involvement of tissues above the maxilla and nasolabial fold and then there is gradual progression to angle of mouth, eyes, eyebrows, ears and neck. The eyes and cheeks of the affected side are sunken and whitening of facial hair and loss of hair (alopecia). The skin overlying the affected areas may become darkly pigmented (hyperpigmentation) in some cases, areas of hyperpigmentation and hypopigmentation. Some patients present a demarcation line between normal and abnormal skin, reminding a big linear scar, known as "*coup de saber*," (Holland and Nocton) as could be noticed in this patient. The deterioration may also affect the tongue, the soft and fleshy part of the roof of the mouth, and the gums (Stone J).

Parry-Romberg syndrome is also accompanied by neurological abnormalities including seizures and episodes of severe facial pain (trigeminal neuralgia).Trauma, viral infections, endocrine disturbances, auto-immunity and heredity are believed to be also associated to the pathogenesis of the disease.The onset of the disease usually begins between the ages of 5 and 15 years. The progression of the atrophy often lasts from 2 to 10 years, and then becomes static. Muscles in the face may undergo atrophy and there may be bone loss in the facial bones. Problems with the retina and optic nerve may occur when the disease surrounds the eye (Stone J)

There is no cure and there are no treatments that can stop the progression of Parry-Romberg syndrome. Reconstructive or microvascular surgery may be needed to repair wasted tissue. The timing of surgical intervention is generally agreed to be the best following exhaustion of the disease course and completion of facial growth. Most surgeons will recommend a waiting period of one or two years before proceeding with reconstruction. Muscle or bone grafts may also be helpful. Other treatment is symptomatic and supportive. The prognosis for individuals with Parry-Romberg syndrome varies. In some cases, the atrophy ends before the entire face is affected. In mild cases, the disorder usually causes no disability other than cosmetic effects.

Discussion

On presentation with the above features which were uncommon the diagnosis was not easy and we had a differential diagnosis of Aplasia cutis, Parry Romberg and

Volume : 5 | Issue : 7 | July 2015 | ISSN - 2249-555X

Goltz syndrome (focal dermal hypoplasia). Focal dermal hypoplasia (FDH) is an uncommon genetic disorder characterized by distinctive skin abnormalities and a wide variety of defects that affect the eyes; teeth; and skeletal, urinary, gastrointestinal, cardiovascular, and central nervous systems. It is usually, but not always, X-linked dominant (lethal in males). About 90% of affected individuals are female. The mnemonic FOCAL can be used to remember some of the key features of this syndrome: female sex; osteopathia striata; coloboma; absent ectodermis-, mesodermis-, and neurodermis-derived elements; and lobster claw deformity. Focal dermal hypoplasia is also known as Goltz syndrome or Goltz-Gorlin syndrome.

With the help of Dermatologist and skin biopsy, it helped us in confirming the diagnosis of Parry Romberg syndrome.

Conclusion

Since it's a rare disorder and the diagnosis can be difficult at times, biopsy helps in the diagnosis. The challenge lies in the management since being a girl child it is cosmetically disfiguring and parents found it very difficult to digest. Good cosmetic surgery helps in reducing the stigma.

Cicatricial alopecia

Linear scleroderma

En coup de sabre lesion

Linear scleroderma

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