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PARRY ROMBERG SYNDROME IN A PEDIATRIC PATIENT

Paediatrics

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ABSTRACT

Parry Romberg Syndrome is a rare disorder of suspected Autoimmune Etiology. First Described by Parry in 1825 and Romberg in 1846. Eulenberg in 1871 termed it as Progressive facial hemi atrophy. The syndrome is characterised by slow atrophy involving one side of face. The affected individuals are normal at birth, with insidious atrophy progression in first two Decades of life.. We Report a unique case of 9 years girl with parry Romberg syndrome with no neurological sequalae but with hypo plastic maxillary bones, malocclusion of teeth, atrophy of tongue hepatosplenomegaly and failure to thrive.

KEYWORDS:

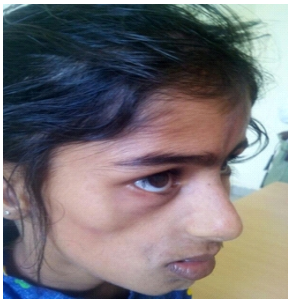
Parry Romberg, facial hemiatrophy, hypoplasia maxilla

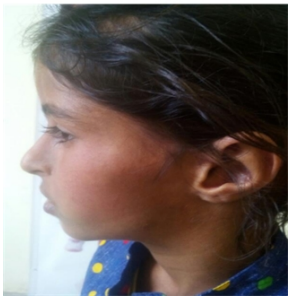
INTRODUCTION:-

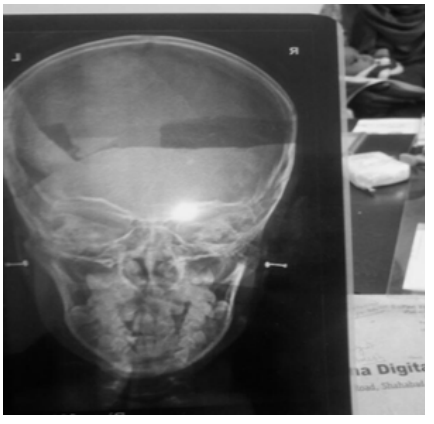
Parry Romberg syndrome is also known as progressive hemifascial atrophy [1.the disease was first describ4ed a by parry in 1815 and Romberg in 1846 [2],[3]. It is characterized by slow and progressive atrophy of skin and subcutaneous tissue , but may extend to muscles and bones of face[4],[5]. The syndrome is characterised by slow atrophy involving one side of face. The affected individuals are normal at birth, with insidious atrophy progression in first two Decades of life. The Disease Remember scleroderma with atrophy of skin, subcutaneous tissue fat, and rarely muscle and Bone. The Progressive of atrophy may extend upto 20 Years ,Following which the patient stabilizes; It is seen commonly in females and associated with neurological ,ocular ,oral deformities like trigeminal neuralgia ,seizures Enopthalmosis ,atrophy of tongue etc.Diagnosis is based on history

CASE REPORT:

A 9 year old girl Presented with complaints of inability to gain weight, short stature and deformity of face. The child was well up to 6 year of age followed by slowly shrinking of Right side of face noticed at 6 years of age and then increasing progressively and now having hollowed appearance of the cheeks. The patient had deformity of upper lips with deviation of lip to right side .She was well growing up to 6 year but not gaining weight and height for last 3 years .Parents complained of short stature as compared to other peers in school and increasing hallowing of right side of face . .No history of trauma. prolonged infection or contact with tuberculosis. No other family sibling was suffering from similar illness. Clinical examination revealed a malnourished girl child with weight 20kg [< 3rd centile] ,Height 120 cm[< 2 S.D].V ital signs were normal .-.Liver 3cm BRSCM,spleen was palpable. She was alert, with no signs of neurological abnormality. Speech and hearing was normal. Millstones were normal for age except malocclusion of teeth noticed at of 8 years age. The face was asymmetric with right sided atrophy .The right maxilla and zygomatic region appeared hypo plastic with shrinking of skin and giving a hollow appearance of cheek on right side [Figure 1] as compared to the normal left side [Figure 2].The Right side of upper lip showed deviation towards the side of contracture. Chin was also deviated to right side . Ears and eyes were normal. Intra oral examination revealed mixed dentition stage with deformity of upper teeth and atrophy of tongue .. Child was investigated. Blood counts, antibody testing for autoimmune possibility, radiography and ultrasonography were done . Antibody testing for antinuclear antibodies ANA was negative. IGA Tissue Transglutaminase antibody was normal .Thyroid profile normal.







Ultrasound abdomen showed liver enlarged span 11CM normal texture . Spleen was enlarged span 8. 2 cm .X-ray of skull including orbit , sinuses and facial bones was done which revealed hypoplastic maxillary bones, small maxillary sinuses ,malocclusion of teeths .Orbits were normal, mastoid bones normal [figure 3]Based on clinical and radiological findings the diagnosis of progressive facial hemi atrophy hence Parry Romberg syndrome was made. Plastic surgeon opinion was taken and she was planned for cosmetic surgery once the disease stabilizes.

DISCUSSION:-

Parry Romberg syndrome is also known as progressive hemifascial atrophy [1.the disease was first describ4ed a by parry in 1815 and Romberg in 1846 [2],[3]. It is characterized by slow and progressive

atrophy of skin and subcutaneous tissue , but may extend to muscles and bones of face[4],[5].The disease is usually involves left side of face but may extend to one side of neck and whole body [5].The disease has a prevalence of 1 in 70000 affecting mainly females [5]but in our case right side was involved .The disease has onset in first decade of life , progresses and then stabilizes[6],[7].The patient in our case has disease onset at 6 years of life and approached to our hospital at 9 years of life .The exact etiology is unknown and various factors have been proposed including autoimmunity, viral infection, peripheral neuropathy, vascular disturbances, increased sympathetic activity causing fascial atrophy [8].the disease can present with ocular, neurological feature like trigeminal neuralgia, epilepsy , focal paresthesia, headache [9]. Imaging findings on CT includes intracranial calcifications, vascular malformations , demyelinations [10]. Treatment aims at correction of fascial deformity after cessation and stabilization of disease process, usually at end of second decade. Plastic surgeon opinion was taken in our case and advised facial augmentation with grafting by 18 years of age.

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