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FOIX-CHAVANY-MARIE SYNDROME AN UNUSUAL PRESENTATION OF HERPES ENCEPHALITIS

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Abstract : FOIX-CHAVANY-MARIE SYNDROME also known as Bilateral Anterior opercular syndrome is a rare cortical form of supranuclear (pseudobulbar) palsy in which there is an Automatic-voluntary dissociation of motor function of lower cranial nerves.There are very few reported cases of this syndrome in childhood, reported as a feature of herpes encephalitis. We report the case of a young girl, who presented with this syndrome as a feature of herpes encephalitis.

Keyword : Herpes encephalitis, Anterior operculum, Pseudobulbar palsy, Automatic-voluntary dissociation

CASE REPORT:

A 7-year- old female child, 4th born to non-consanguineous parents was referred to our casualty as a case of acute central nervous system infection. Her chief complaints were fever, impairment of speech, feeding difficulties, focal seizures involving upperlimbs, face and perioral region and altered sensorium of 2 days duration. There was no significant past medical history, family history or contact history. Her development was normal and scholastic performance was good.

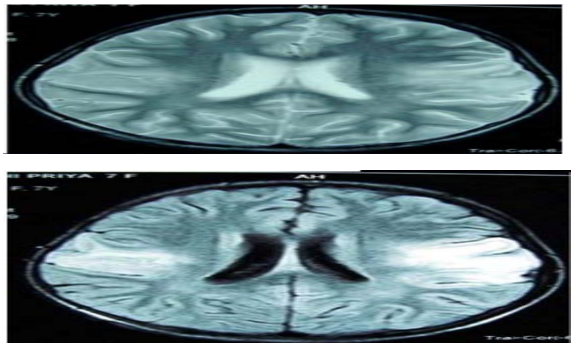
On examination the child was unconscious and pain responsive with focal seizures involving both upperlimbs, face and perioral region. Heart rate -90/minute, Respiratory rate-30/minute, Blood pressure-110/70mm Hg . Examination of other systems revealed no abnormality.

7YR OLD GIRL WITH BILATERAL ANTERIOR OPERCULAR SYNDROM

Investigations revealed leucocytosis with polymorphonuclear predominance. Liver function test, Renal function tests, electrolytes , chest X-ray, Electro-cardiography and Echo- cardiography were within normal limits. Cerebro spinal fluid examination done on 4th day of illness revealed White Blood cells, with lymphocyte predominance , normal sugar and elevated protein (sugar-63, protein-66mg/dl) values.

Herpes Simplex Virus DNA was detected in Cerebro spinal fluid by Polymerase Chain Reaction(Day 4).Magnetic Resonance Imaging of brain revealed hyperintensities involving both temporal lobes and perisylvian region(Left>Right) . Electroencephalography revealed periodic lateralizing epileptiform discharges.

Child was treated with Acyclovir, antibiotics, anti-edema measures, multiple anticonvulsants, and other supportive measures. Child improved gradually over a period of 2 weeks with residual weakness of right upperlimb and pseudobulbar palsy of cortical type with difficulty in opening the mouth voluntarily



T1 AND T2 WEIGHTED MRI IMAGES SHOWING THE AFFECTED AREAS

DISCUSSION:

Magnus in 1837 [1] and is known as Foix-Chavany-Marie syndrome (FCMS) [2] . Anarthria and bilateral central facio-linguovelo -pharyngeo-masticatory paralysis with "automatic voluntary dissociation" are the clinical hallmarks of Foix-Chavany-Marie syndrome (FCMS). Weller et al reviewed 62 cases, which allowed the differentiation of five clinical types of this syndrome: a)the classical and most common form associated with cerebrovascular disease, (b) a subacute form caused by central nervous system infections, (c) a developmental form probably most often related to neuronal migration disorders, (d) a reversible form in children with epilepsy, and (e) a rare type associated with neurodegenerative disorders[3]. Manifestations include volitional paralysis of muscles innervated by cranial nerves 5, 7, 9, 10 and 12 with preserved autonomic and emotional innervations of these muscles. (eg; emotional smiling, laughter, crying, yawning.) It differs from bulbar palsy by preservation of jaw jerk, pharyngeal reflex and by the absence of fasciculation, atrophy and phenomenon of denervation and unlike pseudobulbar palsy the pathological laughter and emotional disturbances.[4]

The typical presentation and differential diagnosis of Foix-Chavany -Marie syndrome

provide important clues to lesion localization in clinical neurology. FCMS is a paretic and not an apraxic disorder and is not characterized by language disturbances. Its clinical features prove divergent corticobulbar pathways for voluntary and automatic motor control of craniofacial muscles. Precise clinico-neuroradiological correlations should facilitate the identification of the structural substrate of "automatic voluntary dissociation" in FCMS[3].

There are very few reported cases of this syndrome in childhood. The reported cases in children occurred following developmental abnormalities, epilepsy and herpes encephalitis. Among these causes, most of the cases in children were reported in association with herpes encephalitis. Van der poel et al, had reported 4 infants with this syndrome associated with herpes encephalitis. Herpes Simplex Virus has a predilection for the opercular area. Herpes Simplex Virus usually presents with fever, altered sensorium and focal neurological deficits. Atypical neurological manifestations have been repeatedly reported in children. Opercular Syndrome is one of the atypical presentation of herpes simplex encephalitis. In some cases of herpes encephalitis, opercular syndrome will manifest as a sequelae also[5]. There is an increasing number of reports describing acute opercular syndrome as one of the intial neurological manifestations of Herpes simplex encephalitis in children[6,7]

The child presented in my case report acquired this syndrome as a feature of herpes encephalitis. Prognosis of this syndrome depends on the etiology. It is very good if it occurs as a result of epilepsy [8]. Here in my case the child is not able to open the mouth for feeding and for talking but she opened the mouth for yawning and crying . The residual weakness of her right upperlimb improved with physiotherapy and she started writing for communicating with her parents for her needs with the language she knew. She was started on feeding through Ryle's tube and discharged with the same after educating the mother. She came for follow-up after a period of 4 months and by this time she started taking the feeds orally with little difficulty and she uttered a few words.

CONCLUSION:

Recognition of the opercular syndrome as a atypical presentation of herpes encephalitis enables early diagnosis and rapid initiation of treatment with acyclovir in these patients, which will improve the clinical outcome.

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