Research Paper Medical Science

Rassmussens Encephalitis in 17 year old Adolescent boy

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ABSTRACT

Rasmussen encephalitis (RE) is a rare but severe immune-mediated brain disorder leading to unilateral hemispheric atrophy,hemiparesis, associated progressive neurological dysfunction and intractable seizures. We report the case of a 17years old adolescent boy with uncrossed hemiplegia and Upper motor neuron type of Facial nerve palsy following birth asphyxia .

KEYWORDS Rasmussen encephalitis,CT imaging

Introduction :

Rasmussen encephalitis (RE) is a rare disease of unknown etiology that causes severe chronic unihemispheric inflammatory disease of the central nervous system mainly in children. It leads to intractable seizures, cognitive decline and progressive neurological deficits in the affected hemisphere.1 Though the etiology of this sporadic disease is unclear, cytotoxic T cell reaction against the neurons was implicated in the pathogenesis.2 Imaging plays a pivotal role in diagnosis by exclusion of other causes and helps towards monitoring the disease progress. Early institution of immunotherapy has been suggested to improve the outcome and alter the natural history of disease.3 Here, we report a Adolescent boy diagnosed with RE based on clinical features, and imaging findings.

Case Report :

An 17 year old adolescent boy presented to Pediatric OPD with unable to hold glass of water and plate for past 5 years and difficulty in combing , and difficulty in walking using right leg. Child is a known case of seizure disorder (complex partial seizures) from 10 years of age and is on antiepileptic medications. Perinatal history of Birth Asphyxia present .Developmental milestones were almost normal except gross motor; standing and walking were delayed .On examination , vitals were stable. In CNS examination, higher motor functions were normal except mild intellectual impairment. Cranial nerve examination revealed right sided UMN type of facial nerve palsy and examination of other cranial nerves were normal. Bulk and power(4/5) of right upper and lower limbs were decreased compared to left and tone was increased. Deep reflexes were exaggerated with ankle clonus and Babinski sign was positive on right side .



Figure no 1 Right upper limb showing significant wasting

Blood investigations were within normal limits done in our hospital.Previously chid was shown in Nimhans hospital done all investigations including EEG and MRI scan which patient attenders lost 3 years back while travelling in bus.

Here again CT imaging was done which showed hemiatrophy of left cerebral hemisphere with dilatation of left lateral ventricle and wallerian degeneration of Descending tracts

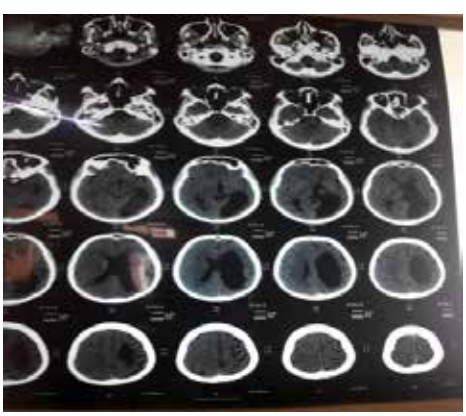


Figure no 2. FdescFF

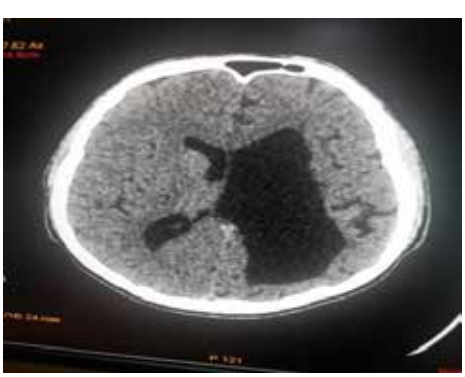


Figure no 3

Hemiatrophy of the left cerebral hemisphere with dilatation of the left lateral ventricle .

Discussion :

Rasmussen encephalitis is a sporadic chronic inflammatory disease of the central nervous system occurring mostly in the pediatric population, first reported by Theodore Rasmussen in 1958. The mean age of presentation 85% is under 10 years, Both sexes are equally affected.1 Our patient was in the same age group. The etiology of RE is unknown, with some earlier studies suggesting the role of viral infections, while others describing it as an autoimmune phenomenon involving antibodies against a protein of glutamate receptor.3,4 According to a recent concept, cytotoxic T cell reaction against the neuron leads to expression of major histocompatibility complex (MHC) class I and apoptotic neuronal death, resulting in progressive deterioration of neurological status.2 No specific etiology could be found in our patient either. Clinically, RE presents as epilepsia partialis continua (EPC) followed by hemiparesis and cognitive impairment, which gradually progresses with the disease activity. Diagnosis of is Rasmussen encephalitis based on characteristic clinical, radiological, and pathological features with more emphasis on clinico-radiological features, as brain biopsy due to its invasive nature is not done in all the cases.5,6 The first stage is nonspecific, called the prodromal stage, manifesting with a relatively low seizure frequency and, rarely, mild hemiparesis . The second phase, called the acute stage with a median duration of 8 months (range: 4-8 months), is characterized by an augmentation in the frequency of seizures, often as EPC, and an increase in the degree of hemiparesis. The final residual stage presents with permanent and stable neurological deficits, mostly severe hemiparesis, and a decreased frequency of seizures.

7,The differential diagnoses for RE include Dyke-Davidoff-Masson syndrome, Sturge-Weber syndrome, hemimegalencephaly, and unihemispheric cerebral vasculitis. Dyke Davidoff Masson syndrome (DDMS) is a set of conditions of different etiologies, leading to unilateral cerebral atrophy with homolateral calvarial hypertrophy and hyperpneumatization of sinuses. The etiology may be classified into congenital and acquired groups. In the congenital variety, cerebral damage usually has a vascular origin. In the acquired type, cerebral insults occur during perinatal period or later and causes include trauma, intracranial hemorrhage, infection and ischemia; and in premature infants, subependymal germinal matrix and intraventricular hemorrhage. As the insult to brain occurs much earlier in intrauterine or perinatal period compared to RE, there is a compensatory overdevelopment of paranasal sinuses and mastoid air cells, ipsilateral calvarial thickening and elevation of the petrous ridge, sphenoid wing and orbital roof.8,Our patient did not present with any of these features.

The Sturge-Weber syndrome is a rare neurocutaneous syndrome characterized by facial capillary and capillary-venous malformations in trigeminal nerve distribution, leptomeningeal venous angiomatosis, seizures, dementia and hemiplegia. Characteristic imaging findings of the cerebral atrophy with gyral or curvilinear calcification (typically described as tram track), enhancing angiomas and ipsilateral enlarged choroid plexus readily differentiate it from RE. Secondary compensatory skull changes to atrophy as in DDMS are also described, but considered as a separate entity

Conclusion :

CT abnormalities of RE evolve from the initial unihemispheric swelling and images show hemiatrophy and dilatation of left cerebral hemisphere and left lateral ventricle . In pediatric unihemispheric progressive cerebral atrophy, Rasmussen encephalitis should be considered in the differential diagnosis, especially if there is no intracranial calcification or contrast enhancement. The radiologist plays an active role as imaging is an important tool for early diagnosis and excluding differential diagnoses, which can modify the progression of disease.

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