

An interesting case of hemiparesis with resistant epilepsy- *Dyke, Davidoff, and Masson syndrome*

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Introduction

Last year a 19 year old boy presented to our epilepsy clinic with a history of right focal motor seizure without secondary generalization, right-side hemiparesis, and mental retardation since 6 month years of age. He was born out of a non-consanguineous marriage with a full-term normal delivery without any antenatal or perinatal complications. He had normal developmental milestones up to 6 moth of age. At 6 moth age he developed fever, vomiting, altered sensorium and convulsion. He was treated in a state medical college hospital as a case of meningoencephalitis of tubercular etiology based on CSF and clinical presentation, and he was discharged subsequently after 1 month on antiepileptic medications. However, he continued to have seizures which exacerbated due to poor compliance and other precipitating factors like sleep deprivation, stress, etc. Besides, the child also had some learning difficulties so he couldn't continue formal education though he developed ability to read, write. By age of 8-9 years his speech became slurred, right face was weak, and had progressive right hemiparesis, for which he was treated conservatively by a pediatrician. Among behavioral problem he has over aggressive behavior. Seizures did not respond to several antiepileptic medications in different combinations. However, the seizure frequency had decreased over the past 6 years, and now seizure occurring only in night with frequency of 1-4 per month. In Initial 3 years the patient was on a two antiepileptic drugs e.g. phenobarbitone and valproate had frequent episode of seizures. Later on because of poorly control seizure, he moved physician to physician and put on various combinations of valproate, phenytoin, carbamazepine, clobazam, levetiracetam. Since last 1 year patient is in regular follow up at our epilepsy clinic and we gradually withdraw clobazam, levetiracetam, valproate, phenytoin and put him on adequate dose of carbamazepine 800 mg daily. Aside this, the patient's hemiparesis had get worse gradually with increased stiffness in the right extremities. His examination did not reveal any neurocutaneous markers. He scored poorly on the Mini-Mental Status Examination (17/30) and had a right homonymous hemianopia with sparing of macula, right sided spastic hemi-paresis with brisk tendon reflexes and extensor

plantar response. He is able to do ADL with minimum support. A magnetic resonance imaging of the brain was afterward done, which discovered near complete atrophy of the left cerebral hemisphere (Fig. 1) along with thickening of the calvarium on the same side (Fig. 2). EEG shows left focal slowing (Fig. 3). We thus kept a diagnosis of DDMS and managed him conservatively with baclofen, physiotherapy and counseling, after which her symptoms improved somewhat.

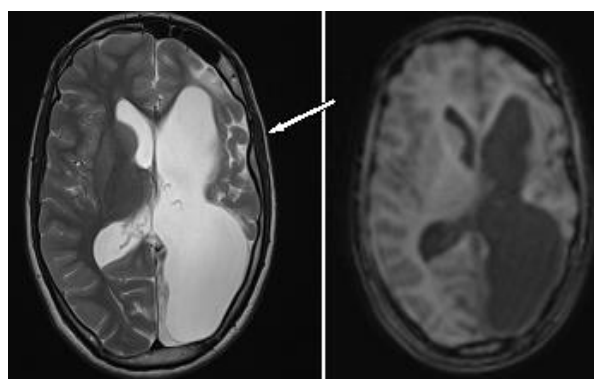


Fig. 1: T2 (right) and T1 (left) weighted axial MRI showing cerebral hemiatrophy on the left side of the brain at the level of the basal ganglia

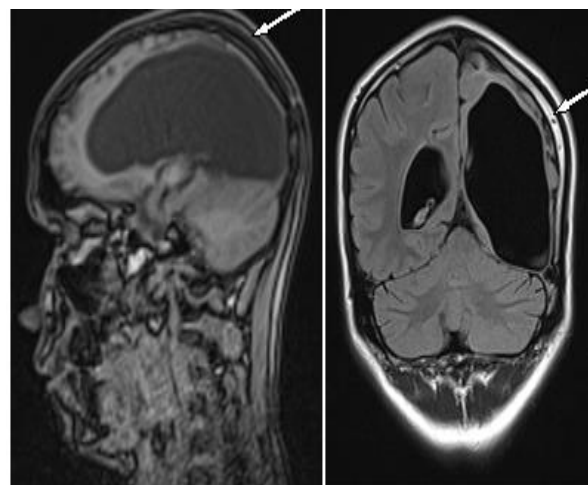


Fig. 2: MRI of the brain showing left hemiatrophy along with calvarial thickening in an sagittal and coronal view T1-weighted sequence (thick white arrow)

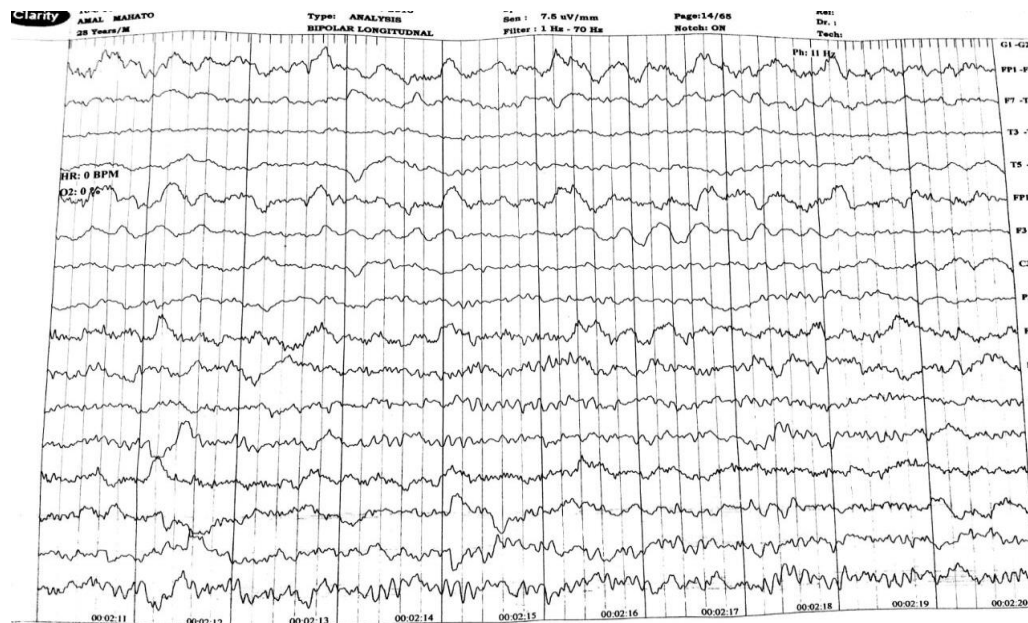


Fig. 3 Awake EEG of patient shows left focal slowing

Discussion

This infrequent condition took its name after the researchers Dyke, Davidoff, and Masson who initially reported the condition way back in 1933. They described skull radiographic changes in 9 patients. Their patients presented with seizures, facial asymmetry, hemiparesis, and mental retardation.⁽¹⁾ Subtotal or diffuse cerebral hemiatrophy is a classical imaging finding. However, unilateral focal atrophy may occasionally be noted in the cerebral peduncles and the thalamic, pontine, crossed cerebellar, and parahippocampal regions. Brain imaging may additionally reveal prominent cortical sulci, dilated lateral ventricles and cisternal space, calvarial thickening, ipsilateral osseous hypertrophy with hyperpneumatization of the sinuses, and an elevated temporal bone.^(1,2) There is no sex predilection, and any side of the brain can be involved, although involvement of the left side and male gender has been shown to be more common in one study.⁽²⁾

There are two identified types of cerebral hemiatrophy. First the infantile or congenital variety results from neonatal or gestational vascular occlusion involving the middle cerebral artery, unilateral cerebral arterial circulation anomalies, coarctation of the mid-aortic arch, or infections, and patients become symptomatic in the perinatal period or infancy. Second one is acquired type results from various causes like birth asphyxia, prolonged febrile seizures, trauma, tumor, infection, ischemia, and hemorrhage.^(3,4) The classical MRI changes of this disease occur only if there is a brain insult earlier than 3 years of age.⁽³⁾

Our understanding of the brain pathology in this syndrome is related to the development of the brain minutely. The formation of the brain sulci starts around

the 14-16 week of conception and finished by the end of the 36 week. Overall, the maximum growth of a head occurs in the early years due to outward pressure of the enlarging brain on the bony skull table which reaches half of its mature size at the end of infancy and three fourths of the mature size by the end of 3 years.⁽⁵⁾ Therefore, only when brain damage is sustained before 3 years of age, other structures overlying the brain grow inward, thus resulting in an increased width of the diploic spaces, enlarged sinuses, and an elevated orbital roof, which are characteristic of this disorder.⁽⁴⁾ The plausible mechanism of cerebral atrophy and the related progressive neurodeficit is hypothesized to be due to several ischemic episodes resulting from different causes, which reduce the production of brain-derived neurotrophic factors, which in turn leads to cerebral atrophy.⁽⁶⁾

This patient had the acquired variety of the disease as the patient's complaints started after an episode of meningoencephalitis at the age of 6 months. There was a progressive increase in spasticity and weakness on the right side. Our patient strikingly had complete left cerebral hemiatrophy, which is atypical for the disease, especially for the acquired variety. Although initially the patient had progressive symptoms in the form of intractable seizures, followed by progressive hemiparesis, which is atypical for this condition, as the classical presentation of this disease itself is an intractable seizure. However, the natural course of this disease in adults has not been established due to the scarcity of adolescent or adult presentation. There have been few pediatric case reports described where, despite progression of the disease in the form of hemiatrophy of the brain and hemiparesis, the dramatically reduced seizure frequency.^(6,7)

Homonymous hemianopia, as seen in our case, has been reported previously in scientific studies.⁽⁸⁾ MRI findings were classical in our case and have been described earlier.^(2,8)

The differential diagnosis of this syndrome includes Rasmussen encephalitis, Silver-Russell syndrome, and Fishman syndrome. Most of these, however, can be differentiated by performing a thorough clinical examination and by neuroimaging.^(3,9) Rasmussen encephalitis, a chronic progressive immune-mediated disorder of children between 6 and 8 years of age, also presents with intractable focal epilepsy and cognitive defects with similar imaging findings of hemispheric atrophy, but calvarial changes are not seen.⁽¹⁰⁾ Silver-Russell syndrome is characterized by the classical facial phenotype (triangular face, small pointed chin, broad forehead, and thin wide mouth), poor growth with delayed bone age, clinodactyly, hemihypertrophy with normal head circumference, and normal intelligence.⁽¹¹⁾ Fishman syndrome or encephalocraniocutaneous lipomatosis is a rare neurocutaneous syndrome including unilateral cranial lipoma with lipodermoid of the eye, which presents usually with seizures. Neuroimaging, however, shows a calcified cortex and hemiatrophy.⁽¹²⁾

Conclusion

For DDMS cases presenting in early childhood, refractory seizures remain the usual concern. Accordingly, hemispherectomy is the treatment of choice with a variable success rate. However, if the presentation is late as in our case and if seizures are under reasonably control, the patient can be reserved on antiepileptic medications instead of surgery, along with supportive therapy including physiotherapy, speech therapy, and occupational therapy. Further longitudinal studies are required to ascertain the natural course of this syndrome especially in an adult population, which would help in planning strategies regarding the time and nature of interventions and management accordingly.

Statement of Ethics

The subject of this case report gave his informed consent for the publication of this article. We did follow treatment protocol for epilepsy and symptomatic therapy for spastic hemiparesis.

Disclosure Statement

The authors declare that they have no conflicts of interest.

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