Dyke-Davidoff-Masson Syndrome

An 8-year-old boy presented with uncontrolled seizures. He was born as the second child to non-consanguineous parents. He had significant perinatal asphyxia in the newborn period. He had developmental delay since infancy and was noticed to have right-sided tonic clonic seizures since the age of 3 years, which was uncontrolled on phenytoin. He was left-handed and had concerns in the domains of speech, learning, behaviour and sleep.

On examination, he had microcephaly. The bilateral carotid pulsations were normal with no bruit. The neurological examination revealed right upper motor neuron facial palsy with right hemiparesis. The developmental evaluation showed that he was functioning at around 3 years of age. He had needs in the domains of learning, speech and behaviour. He was hyperkinetic, anger prone and had episodes of temper tantrums. He spoke in simple sentences and was able to communicate his needs.

His blood investigations were normal. An electro-encephalogram revealed left hemisphere dysfunction. The transaxial sections of magnetic resonance imaging (MRI) brain are shown in Figures 1 and 2. What is the radiological diagnosis?

a. Sturge-Weber syndrome
b. Unilateral megalencephaly
c. Dyke-Davidoff-Masson syndrome
d. Silver syndrome
e. Rasmussen encephalitis

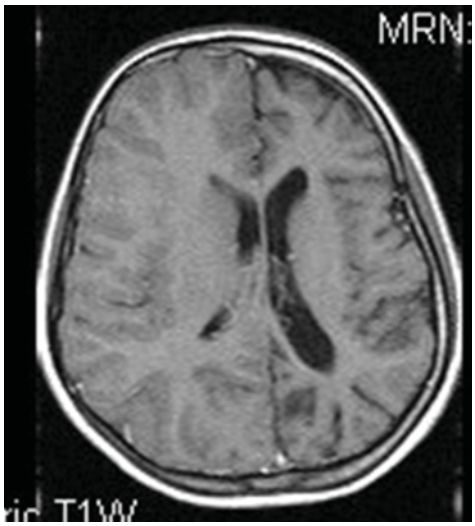

Fig. 1. The magnetic resonance imaging of the brain shows the characteristic picture on T1 weighted axial image.

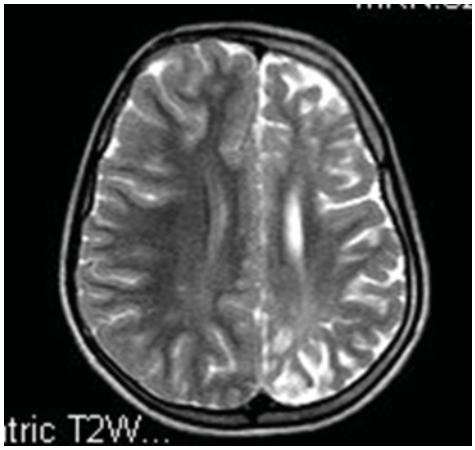

Fig. 2. The T2 weighted axial image on magnetic resonance imaging also shows the typical parenchymal and osseous changes.

Discussion

The MRI brain shows left frontal and parietal lobe atrophy with dilatation of left lateral ventricle, thinning of the body of corpus callosum and thickening of left frontal cranium and elevation of left orbital roof, consistent with the diagnosis of Dyke-Davidoff-Masson syndrome (DDMS).

DDMS refers to the condition where cerebral hemiatrophy occurs due to an insult to the developing brain in the foetal or early childhood period. It is characterised by asymmetry of cerebral hemispheric growth with atrophy of one side and midline shift, ipsilateral osseous hypertrophy with hyperpneumatisation of sinuses – mainly frontal and mastoid air cells with contralateral paresis.[1]

The developing brain, in the foetal life, infancy and early childhood, presses outwards on the bony skull table to give the gradual increase in head size and shape. When the brain fails to grow properly, the other structures grow inward, resulting in increased width of diploic space, enlargement of the frontal sinus and elevated orbital roof.

There are 2 types – infantile (congenital) and acquired. The infantile variety results from various aetiology such as neonatal or gestational vascular occlusion involving the middle cerebral artery and unilateral cerebral arterial circulation anomalies. The main causes of acquired type are tumour, infection, ischaemia and haemorrhage.[2]

The major clinical manifestations are hemiparesis, uncontrolled seizures and cognitive departure. The clinical

Answer: c

presentation varies depending upon the extent of damage to the brain and presents as varying degrees of hemiatrophy, hemiplegia, facial paresis, seizures, cognitive departure, language defect and learning disorder. There was no relationship between the parenchymal changes and the time after onset of the disease.[3] Prognosis is better if the onset is after 2 years of age and the seizures are controlled.[1] Hemispherectomy is a treatment option for intractable seizures.

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