A child with cerebral hemiatrophy: a rare case report of Dyke-Davidoff-Masson syndrome

Clinical History:

A 12-year-old female child was brought to our Outpatient Department with history of epilepsy who was taking antiepileptic drugs. There was history of vacuum delivery at birth without significant family history. Vitals were normal. Examination of the central nervous system revealed hemiparesis on the right side.

Imaging Findings:

Computed Tomography (CT) suggested unilateral hemiatrophy of left cerebral hemisphere with dilated left lateral ventricle and ipsilateral prominent sulci. There is atrophy of left side cerebral peduncle of midbrain, pons and medulla oblongata – Wallerian degeneration of brain stem. Subtle increased thickening of left hemicranium noted. The ipsilateral frontal sinus appears enlarged.

Magnetic resonance imaging (MRI) showed unilateral diffuse atrophy of the left cerebral hemisphere with dilatation of ipsilateral lateral ventricle and ipsilateral sulcal prominence. There was no midline shift. Thickening of the left hemicranium noted, measuring 3.5 mm on the right side and 7.5 mm on the left temporal lobes. Atrophied parts of left sided of midbrain, pons and medulla oblongata also noted clearly. There is T2 and FLAIR hyperintense signal in the visualized white matter of left hemisphere demonstrating gray-white matter loss with asymmetry of left sided basal ganglia. Cerebellar vermis and hemispheres appear normal.

Discussion:

Dyke-Davidoff-Masson syndrome (DDMS) is a rare disease first described in 1933 by Dyke et al who reported skull radiographic findings and pneumatoencephalographic changes in 9 patients [1]. DDMS is classified as congenital and acquired form which is usually associated with brain damage and is diagnosed during early childhood and postnatal life [2]. Only few adult case reports have been published [3]. Congenital abnormalities, infarctions, vascular malformations, infections and vascular occlusion are prenatal causes whereas traumatic birth history, hypoxia, intracranial haemorrhage, tumours, infections are causes of peri and postnatal causes [4]. Our patient is a young adult and the reason of the disease is thought to be traumatic birth history via vacuum.

Contralateral hemiparesis, hemiplegia, mental retardation, facial asymmetry, epilepsy, language disorders, facial asymmetry are the clinical presentations of DDMS [5]. Computed tomography (CT) and magnetic resonance imaging (MRI) findings are the imaging modalities for its diagnosis[6, 7].
Unilateral cerebral hemiatrophy with ipsilateral compensatory thickening of adjacent bone and hyperpneumatization of sinuses are seen on CT [6]. MRI also shows similar changes along with parenchymal signal changes due to gliosis and porencephalic cysts [7]. MRI also demonstrates the gray-white matter loss with hyperintensities on T2-weighted images (diffuse cortical and subcortical atrophy) and asymmetry of the basal ganglia [8]. MRI can clearly differentiate between congenital and acquired types of DDMS.

Midline shift of the structures towards the side of the disease is typically seen in congenital form. This helps to differentiate the cerebral atrophy occurring in early life. The atrophied hemisphere will show prominent sulcal spaces in acquired form which occurs after the end of sulcation, as in our case [8].

Ipsilateral hypertrophy of calvaria, sinus enlargement, elevations of greater wing of sphenoid, petrous ridge, ipsilateral falcine displacement are common findings in those cases which develops early in life (first 2 years of life) which are the compensatory mechanisms for the hypoplastic cerebral hemisphere [9].

Hemimegalencephaly, Sturge-Weber syndrome and Rasmussen encephalitis are the common differential diagnoses [2]. Rasmussen encephalitis doesn’t show any calvarial changes whereas Sturge Weber syndrome is characterized by typical pattern of cortical calcifications and enhancing pial angiomas [10].

There is only symptomatic treatment for epilepsy, hemiplegia, paresis and language disorder. Better prognosis is seen in those cases where hemiparesis had occurred after the age of 2 years and without repetitive seizures. Hemispherectomy is advised for children with intractable disabling and hemiplegia with a success rate of 85% [10, 11].

Statement of obtained patient consent
Written informed patient consent for publication from the guardian was taken.

Differential Diagnosis List: Dyke-Davidoff-Masson syndrome, Rasmussen encephalitis, Sturge Weber syndrome, Hemimegalencephaly, Fishman syndrome

Final Diagnosis: Dyke-Davidoff-Masson syndrome

References:


Description: Computed Tomography (CT) axial section showing diffuse left cerebral atrophy with prominent sulcal spaces and dilated left lateral ventricle. Origin: Department of Radiology, Nepal Medical College Teaching Hospital, Attarkhel, Jorpati, Nepal, 2021.
Description: Computed Tomography (CT) coronal section showing diffuse left cerebral atrophy with dilated left lateral ventricle

Origin: Department of Radiology, Nepal Medical College Teaching Hospital, Attarkhel, Jorpati, Nepal, 2021
Description: T1 weighted MRI image of brain axial section shows atrophy of unilateral left cerebral hemisphere with prominent extraaxial CSF spaces and thickened left calvaria. Origin: Department of Radiology, Nepal Medical College Teaching Hospital, Attarkhel, Jorpati, Nepal, 2021
Description: T1-weighted MRI image of brain axial section shows unilateral cerebral atrophy with associated atrophy of Internal capsule and lateral ventricle dilatation. Origin: Department of Radiology, Nepal Medical College Teaching Hospital, Attarkhel, Jorpati, Nepal, 2021.
Description: T1 weighted MRI of brain axial section showing atrophied left pontine region. Origin: Department of Radiology, Nepal Medical College Teaching Hospital, Attarkhel, Jorpati, Nepal, 2021.
Description: T2 weighted MRI image of brain axial section showing atrophy of left cerebral peduncle with prominent extraaxial CSF spaces and thickened left calvaria. Origin: Department of Radiology, Nepal Medical College Teaching Hospital, Attarkhel, Jorpati, Nepal, 2021.
Figure 7

Description: T2-weighted MRI image of brain axial section shows atrophy of left cerebral hemispheres with associated ipsilateral white matter gliosis and left lateral ventricle dilatation and thickened left calvaria. Origin: Department of Radiology, Nepal Medical College Teaching Hospital, Attarkhel, Jorpati, Nepal, 2021.
Description: T2-weighted MRI image of brain coronal section shows atrophy of left cerebral hemispheres with associated ipsilateral white matter gliosis and left lateral ventricle dilatation.

Origin: Department of Radiology, Nepal Medical College Teaching Hospital, Attarkhel, Jorpati, Nepal, 2021
Description: FLAIR sequence of MRI image of brain coronal section shows diffuse atrophy of left cerebral hemispheres with associated ipsilateral white matter gliosis and left lateral ventricle dilatation

Origin: Department of Radiology, Nepal Medical College Teaching Hospital, Attarkhel, Jorpati, Nepal, 2021