

Dyke–Davidoff–Masson Syndrome: A Case Report

S.Choayb, H.Adil, N.Allali, L.Chat and S.El Haddad

ARTICLE INFO

Article history:

Received: 7 April 2021;

 Received in revised form:
 01 June 2021;

Accepted: 11 June 2021;

Keywords

 Hemicerebral Atrophy,
 Dyke–Davidoff–Masson
 Syndrome,
 Calvarial Thickening.

ABSTRACT

Dyke-Davidoff-Masson syndrome is a rare entity characterized by hemi cerebral atrophy/hypoplasia secondary to brain insult in fetal or early childhood period. We present a case of a 10-month-old girl, presenting since 5 months right-sided body weakness and generalized tonic-clonic seizures. Brain MRI revealed hemicerebral atrophy of the left hemisphere with dilated ventricles on the same side, cerebral infarction, elevation of the sphenoid wing and petrous temporal bone, and slight calvaria hypertrophy.

© 2021 Elixir All rights reserved.

Case Presentation

We present a case of a 10-month-old girl, presenting since 5 months decreased movement of the right upper and lower limbs and generalized tonic-clonic seizures.

Imaging Findings

Imaging revealed hemicerebral atrophy of the left hemisphere with dilated ventricles on the same side, cerebral infarction, elevation of the sphenoid wing and petrous temporal bone, and slight calvaria hypertrophy. These findings were consistent with Dyke-Davidoff-Masson syndrome.

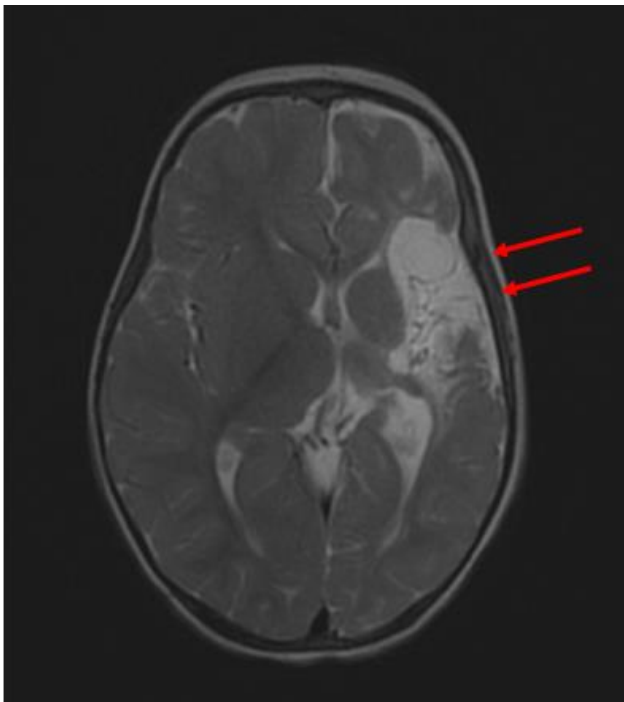


Figure 1. axial T2WI showing hemicerebral atrophy of the left hemisphere and enlarged ipsilateral ventricle with calvaria thickening (arrows).

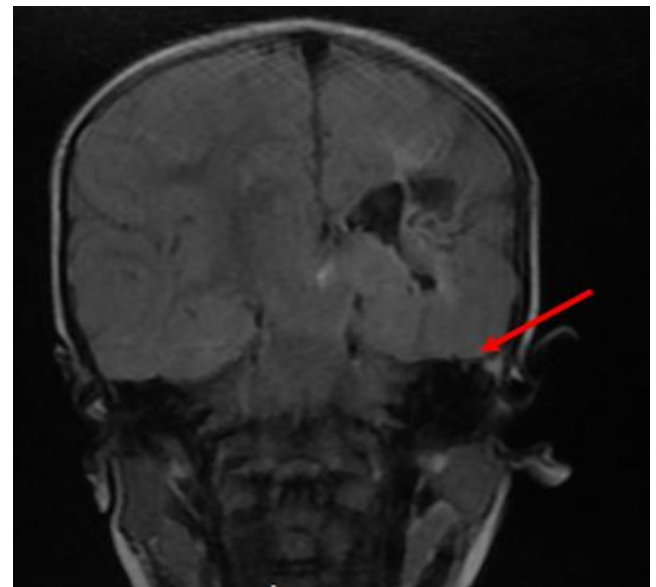


Figure 2. Coronal T2 Flair showing elevation of the sphenoid wing and petrous temporal bone (arrow).

Discussion

Dyke–Davidoff–Masson syndrome (DDSM) is a rare condition resulting from brain insult during fetal life, neonatal period, or early childhood. It was first described in 1933 by Dyke, Davidoff, and Masson on plain radiography and pneumoencephalograms in a series of nine patients with cranial asymmetry and hemiplegia. [1]

Prenatal causes include vascular malformations, intracranial infections, cerebral infarct, and vascular occlusion involving primarily the middle cerebral artery. Peri and postnatal causes include hypoxia, birth trauma, intracranial hemorrhage, tumors, infections, and prolonged febrile seizures. [2]

The clinical presentation depends on the importance of brain injury and includes seizure, contralateral hemiparesis, facial asymmetry, and cognitive disabilities. Patients with

sensory symptoms and psychiatric disorders like schizophrenia can be seen. Mental retardation is not seen in all cases and seizures can appear months to years after hemiplegia. [3][4]

MRI and CT are the gold standard imaging modalities in the diagnosis of DDSM. The typical features include cerebral hemiatrophy/hypoplasia with prominent cortical sulci, ipsilateral ventricular dilatation, interhemispheric fissure is often displaced across the midline, compensatory hypertrophy of the skull with enlargement of frontal sinus, elevation of the greater wing of the sphenoid, and petrous ridge. These findings are more obvious as the patient ages. If brain insult occurs during the fetal period or before age of 3, compensatory calvarial involvement can be seen.

Left-sided hemiatrophy is more common. Other imaging features can include atrophy of the ipsilateral cerebral peduncle and the contralateral cerebellum (secondary to crossed or diffuse cerebellar diaschisis). [2][3][4]

Shen et al described three patterns on MRI: pattern I correspond to diffuse cortical and subcortical atrophy, pattern II corresponds to diffuse cortical atrophy associated with porencephalic cysts and pattern III corresponds to previous infarction with gliosis in middle cerebral artery territory. [5]

The hemisphere involved shows hypoperfusion and decreased metabolic activity using positron emission tomography. [6]

Therapeutic management consists of controlling the seizures associated with domiciliary physiotherapy, occupational, and speech therapy. Hemispherectomy is indicated in patients with hemiplegia and refractory epilepsy.

Prognosis is poor if hemiparesis emerges before 2 years of age and if the seizures are recurrent or prolonged. [4] [6]

Final Diagnosis

Dyke–Davidoff–Masson syndrome

Differential Diagnosis List

Dyke–Davidoff–Masson syndrome

Sturge-Weber syndrome

Rasmussen encephalitis

References

- [1] R.Verma and R.Sahu, “Dyke-Davidoff-Masson syndrome,” pp. 1–2, 2012.
- [2] A. Kumar, B. Rehan, and I. A. Panhwar, “Case report dyke-davidoff-masson syndrome secondary to vascular compromise : a rare cause of cerebral hemiatrophy,” vol. 30, no. June, pp. 150–152, 2020.
- [3] A.Y.Jr, M.Saim, H.Maqsood, S. Younus, and M. H. Raza, “Dyke-Davidoff-Masson Syndrome : A Case Report and Review of Literature Dyke-Davidoff-Masson Syndrome : A Case Report and Review of Literature,” no. December, 2020.
- [4] R.Arora and J.Y. Rani, “Dyke-Davidoff-Masson syndrome : imaging features with illustration of two cases,” vol. 5, no. 3, pp. 469–471, 2015.
- [5] P. Chaudhary, A. Agrawal, A. Malhotra, and A. Bangari, “Case Report,” vol. 4, no. 4, pp. 626–628, 2017.
- [6] E.Go, M.Beyhan, and R.Sade, “Radiological imaging findings of Dyke – Davidoff – Masson syndrome,” 2017.