

Dyke-Davidoff Masson Syndrome

Dyke-Davidoff Masson Sendromu

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To the Editor,

Dyke-Davidoff Masson Syndrome (DDMS), was first described by Dyke, Davidoff and Masson in 1933 (1). DDMS is characterized by seizures, facial asymmetry, hemiplegia or hemiparesis and neuropsychiatric symptoms. The findings are due to cerebral injury which occurs in utero or early in life (2). The clinical findings may be variable depending on the extent of brain injury. In this letter, we presented a case of DDMS with typical imaging findings.

A 23-year-old female patient presented with seizure to our hospital. She had been following for 10 years and seizures were controlled with antiepileptic drugs. For the last 2 months, seizures was uncontrolled. The laboratory test showed anemia, the others were normal. Magnetic resonance imaging (MRI) was performed because of the uncontrolled seizures. The MR images showed the features of left cerebral atrophy, ipsilateral calvarial thickening, left frontal sinus enlargement and hyperaeration, and left ventricular enlargement which were characteristic of DDMS (Fig 1a, b).

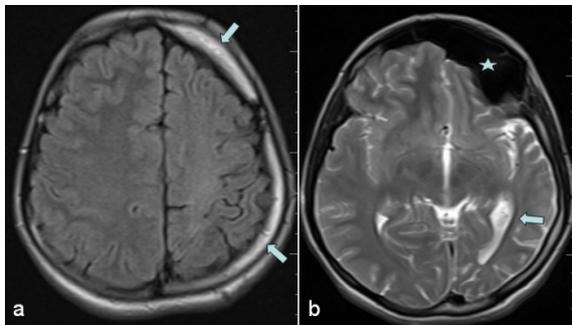


Figure 1. (a) Axial FLAIR image revealed atrophy in left cerebral hemisphere and calvarial thickening (arrow), (b) axial T2-weighted MR image revealed the enlargement and hyperaeration of left frontal sinus (asterisk), and mild ventricular enlargement (arrow).

Hypoplasia of the left middle cerebral artery was also detected on MRI (Fig 2). The diagnosis of DDMS was made with these MRI findings.

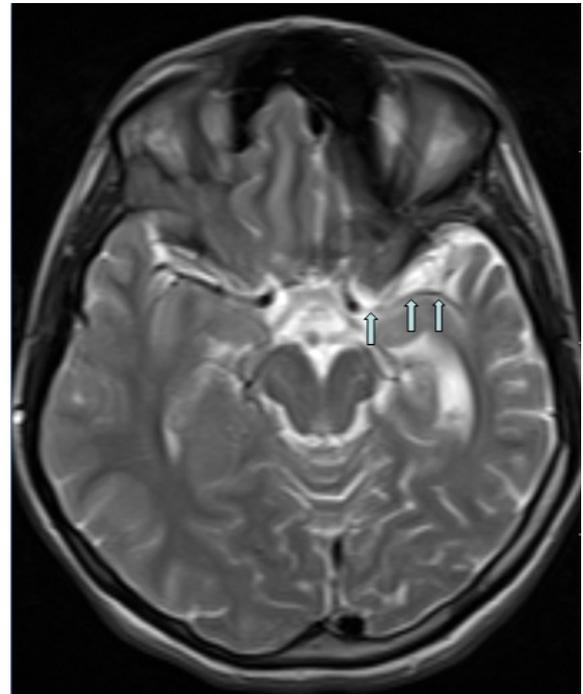


Figure 2. Axial T2-weighted MR image showed the hypoplasia of left middle cerebral artery (arrows).

The pathogenesis of DDMS is thought to originate from a cerebral insult. Causes in the prenatal and perinatal period are congenital malformation, infection, hypoxia, anoxia, birth trauma and intracranial hemorrhage. Postnatal causes are trauma, infection, tumor and seizures (2, 3). The compensatory changes like sinus enlargement and calvarial thickening occurs due to a relative vacuum created by the atrophic brain. Congenital and acquired presentations of DDMS are recognized. In the congenital type structural abnormalities of cerebral arteries, especially middle cerebral artery, have been described as in our case and considered of cerebral atrophy. Few patients

with DDMS have been diagnosed during infancy (4).

The classical clinical presentation includes seizure, facial asymmetry, hemiplegia or hemiparesis and mental retardation. The clinical findings may be of variable degree according to the extent of brain injury.

Plain radiograph of skull may show hyperaeration of paranasal sinuses and calvarial thickening. Computed tomography and MRI findings are characteristic and shows cerebral hemiatrophy, calvarial thickening, enlargement of ipsilateral ventricle and paranasal sinuses. Angiographic examination may show vascular abnormalities (5).

In conclusion, DDMS must be ruled out in differential diagnosis of cerebral hemiatrophy. MRI is a useful method in the analysis of cerebral hemiatrophy and associated abnormalities in such patients. The appropriate clinical history and radiologic findings provide the correct diagnosis.

References

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