

# Dyke - Davidoff - Masson Syndrome may be Confused With Hydrocephalus

## *Dyke - Davidoff - Masson Sendromu Hidrosefali ile Karışabilir*

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### ÖZET

Dyke-Davidoff-Masson (serebral hemiatrofi) sendromu tanısı mevcut dört vakayı sunuyoruz. İlk vaka hidrosefali ile karışabilecek serebral hemiatrofinin ciddi bir formudur. Diğer daha az ciddi serebral hemiatrofisi mevcut üç hastanın ise hidrosefali ile benzer yönü bulunmamaktadır. Sonuçta ciddi derecede serebral hemiatrofisi ve ventriküler genişlemesi var olan vakalar hidrosefali ile karışabilir.

**Anahtar Sözcükler:** Dyke-Davidoff-Masson sendromu, hidrosefali, hemiatrofi.

### SUMMARY

We reported four cases of Dyke-Davidoff-Masson syndrome (cerebral hemiatrophy). The first case is a severe form of cerebral hemiatrophy which may be confused with hydrocephalus. The other three are mild cases which are not so similar with hydrocephalus. In conclusion, the cases with severe cerebral hemiatrophy and ventricular enlargement may be confused with hydrocephalus.

**Key Words:** Dyke-Davidoff-Masson syndrome, hydrocephalus, hemiatrophy

Dyke-Davidoff-Masson syndrome (cerebral hemiatrophy) is a rare condition characterized clinically by facial asymmetry, seizures, contralateral hemiplegia or hemiparesis, and mental retardation (1,2,3). These findings may occur early in life or in utero. Radiologically, magnetic resonance (MR) and computed tomography (CT) demonstrate the parenchymal abnormalities of unilateral loss of cerebral volume and compensatory bone alterations in the calvarium, such as thickening, hyperpneumatization of the paranasal sinuses and mastoid cells as well as elevation of the petrous ridge and greater wing of the sphenoid bone (2,3). Also, there is unilateral ventricular enlargement depending on cerebral atrophy (2,3). Dyke-Davidoff-Masson syndrome may be confused with hydrocephalus because of this radiological feature. In this study we present four cases of Dyke-Davidoff-Masson, one of which shows similarity with hydrocephalus.

## CASE REPORTS

**CASE 1:** An 8-year-old male was evaluated for right hemiparesis and seizures. Presented with locomotor and sensorimotor deficits at the age of eleven month when walking had begun. In examination the only finding was right hemiparesis. MR imaging revealed atrophy at left cerebral hemisphere, mesencephalon, pons and calvarial hypertrophy, hyperpneumatization of the paranasal sinuses with ventricular enlargement on the same side. He continued to visit our department after discharge.

**CASE 2:** A 28-year-old female who suffered from recurrent seizures since the age of six had right facial atrophy detected in physical examination. In neurological examination slight mental retardation and left hemiparesis existed. Brain MRI demonstrated atrophy of the pons, mesencephalon and the right cerebral hemisphere, thickening of the calvarium at the same side and dilatation of the calvarial sinuses with enlargement of the ventricles at right. The patient's seizures underwent control by application of antiepileptic therapy.

**CASE 3:** The present patient who was a 36 year old male suffered from hypoesthesia and seizures. He had a history of dystocia and right hemiparesis in neurological examination. Brain MRI demonstrated mild atrophy of the left cerebral hemisphere and enlarge-

ment of extra-axial, ipsilateral, lateral ventricles. This patient received antiepileptic therapy and his seizures underwent control.

**CASE 4:** A 22 year-old male was evaluated for recurrent seizures persisting since the age of fourteen. She had right hemiparesis in neurological examination. Brain MRI revealed atrophy of the left cerebral hemisphere, medulla oblongata, pons and mesencephalon with enlargement of left lateral ventricle. She was discharged from the hospital after antiepileptic therapy.

## DISCUSSION

Cerebral hemiatrophy is not frequently encountered in clinical practice. When this develops early in life (during the first two years), certain cranial changes like homolateral hypertrophy of the skull and sinuses occur. The compensatory cranial changes occur to take up the relative vacuum created by the hypoplastic cerebrum (2,3). The classical clinical presentation includes seizures, facial asymmetry, contralateral hemiplegia or hemiparesis and mental retardation (1-4). Imaging studies show unilateral atrophy of the cerebral hemisphere with ipsilateral shift of the ventricles. The sulci on the involved side are wide and often replaced by gliotic brain tissue (1-5). The ventricles at the same side with the atrophic hemisphere may seem dilated. In the cases, presented with severe ventricular enlargement, this imaging finding may cause difficulties in differential diagnosis with monoventricular hydrocephalus as in our first case which was thought as a monoventricular hydrocephalus. We were able to diagnose the disease after a further evaluation. Both two pathologies may have the same clinical aspects like contralateral hemiplegia or hemiparesis and mental retardation. Also similar etiologies may exist as prenatal, perinatal, postnatal trauma and infections in this condition. Although MRI and CT imaging findings may seem similar at first sight, many differences may be detected by a careful evaluation. Atrophic sulci of only one hemisphere, atrophic parenchyma, shift on the same side and compensatory changes are the findings of Dyke-Davidoff-Masson syndrome which are important in differential diagnosis. Also parietooccipital diameter is generally normal.

In contrast to the traditionally emphasized features of Dyke-Davidoff-Masson syndrome, in hydrocephalus

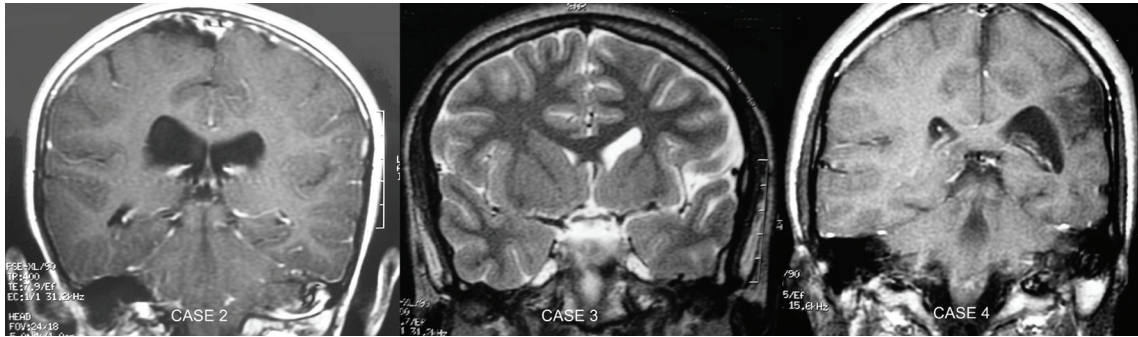


Figure 1A: CT imaging showing right calvarial hypertrophy and hyperpneumatization of the paranasal sinuses in first case. 1B: MR imaging showing atrophy of the right cerebral hemisphere and severe ventricular enlargement on the same side in first case.

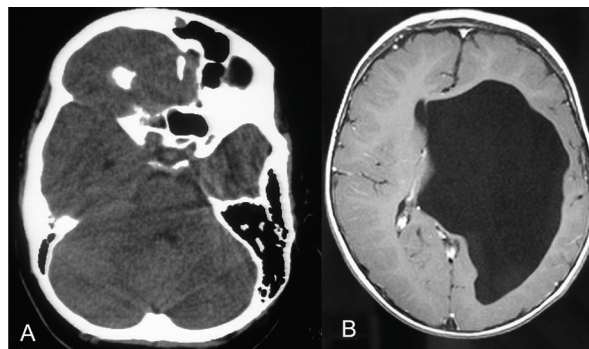


Figure 2: Imaging studies showing unilateral atrophy of the cerebral hemisphere with ventricular enlargement on the same side in other cases.

there is flattening of sulci, periventricular CSF transmission, shift onto the opposite side and increased head's circumference with wide and tight fontanels in pediatric patients. Intraventricular CSF pressure detection and functional MRI may be further methods that are important in differential diagnosis.

The risk of confusion in differential diagnosis with hydrocephalus is not common when the ventricular enlargement is less as in our 2nd and 3rd cases.

In conclusion, the diagnosis of Dyke-Davidoff-Masson syndrome with severe cerebral hemiatrophy and ventricular enlargement may be confused with hydrocephalus. For this reason, history, examination, CT and MRI should be evaluated carefully. Differential diagnosis of Dyke-Davidoff-Masson syndrome requires critical assessment. It may be confused with hydrocephalus and as two different syndromes requiring different treatments a clear identification is needed.

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