

Dyke-Davidoff-Masson Syndrome with Crossed Cerebellar Atrophy

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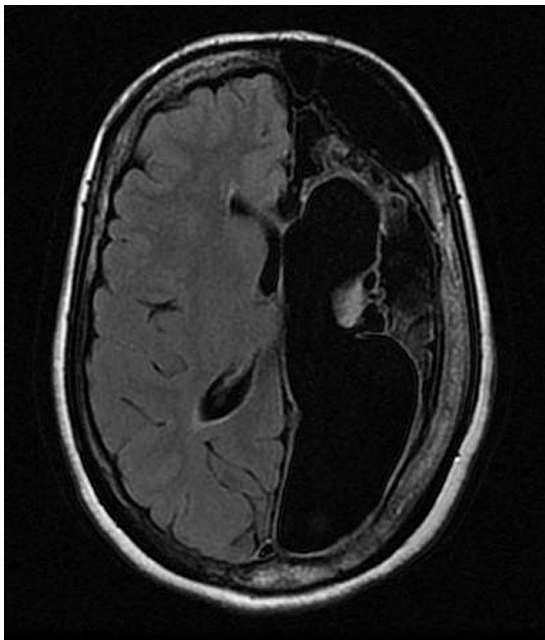


Figure 1. A fluid-attenuated inversion recovery (FLAIR) axial MRI scan reveals left cerebral encephalomalacia with ipsilateral cranial vault thickening, enlargement of the frontal sinuses, and hyperpneumatization.

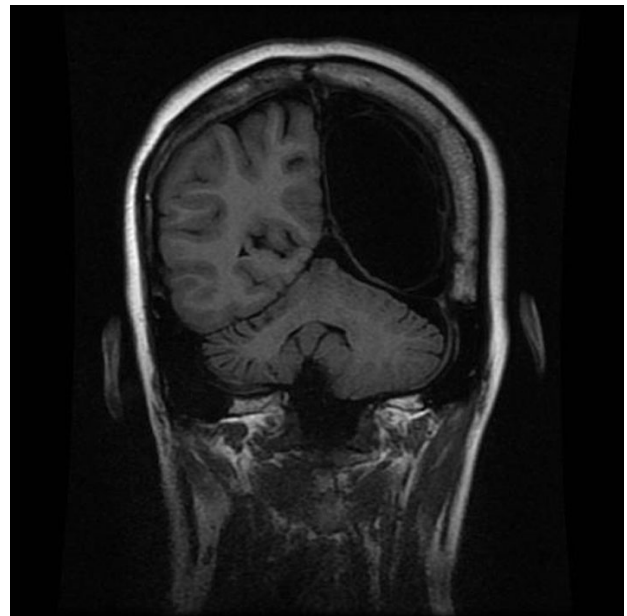


Figure 2. A T1-weighted coronal MRI scan shows crossed right cerebellar atrophy with relative preservation of the middle cerebellar peduncle.

A 52-year-old woman was healthy until the age of 5 years when she suffered from a febrile illness with convulsions. Subsequently, she developed shortness of her right upper and lower extremities with weakness. She was able to function as a housewife after marriage with only mild cognitive impairment. She had no history of epilepsy after the above-mentioned febrile episode.

A neurological examination revealed facial asymmetry and right spastic hemiparesis with prominent atrophy. Brain magnetic resonance imaging demonstrated left cerebral encephalomalacia with ipsilateral cranial vault thickening, enlargement of the frontal sinuses, and hyperpneumatization. There was right cerebellar atrophy with relative sparing of the middle cerebellar

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peduncle.

The Dyke-Davidoff-Masson syndrome (DDMS) was first reported in 1933 in a series of nine patients with hemiplegia who had cerebral hemiatrophy and ipsilateral hypertrophy of the calvarium and sinuses noted on skull X-ray. Clinically, it is usually characterized by facial asymmetry, contralateral hemiparesis or hemiplegia, and epilepsy or mental retardation. The neuroimaging findings are characterized by parenchymal loss in one cerebral hemisphere with ipsilateral compensatory calvarial thickening, hyperpneumatization of the paranasal sinuses, and hypoplasia of the middle-frontal cranial fossa. The causes of cerebral hemiatrophy are diverse and include trauma, infection, ischemia, hemorrhage, etc. It is generally considered that DDMS results from brain damage occurring in utero or early childhood, before maturation of the skull bones is completed ⁽¹⁾.

Dyke-Davidoff-Masson syndrome associated with crossed cerebellar atrophy (CCA) has been previously described. The underlying pathogenesis of CCA may involve damage to the corticopontocerebellar tract or centrolobular cerebellar sclerosis due to prolonged seizure effects ⁽²⁾. A survey for CCA in patients with uni-

lateral precocious destructive brain insults suggested that the extent of the supratentorial lesion and the antecedent of status epilepticus but not recurrent seizures are related to the development of CCA ⁽³⁾.

In conclusion, we report a typical case of Dyke-Davidoff-Masson syndrome accompanied by crossed cerebellar atrophy with relative preservation of the middle cerebellar peduncle in a patient who suffered from febrile illness with convulsions at the age of 5 years without a post-infectious event of epilepsy.

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