

DYKE-DAVIDOFF-MASSON SYNDROME

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Key-word: Brain, atrophy

Background: A 9-year-old boy was referred to the emergency department with seizures. Past medical history revealed an anoxic birth and that the boy had difficulty in learning. Physical examination revealed right-sided hemiparesis. Imaging findings.

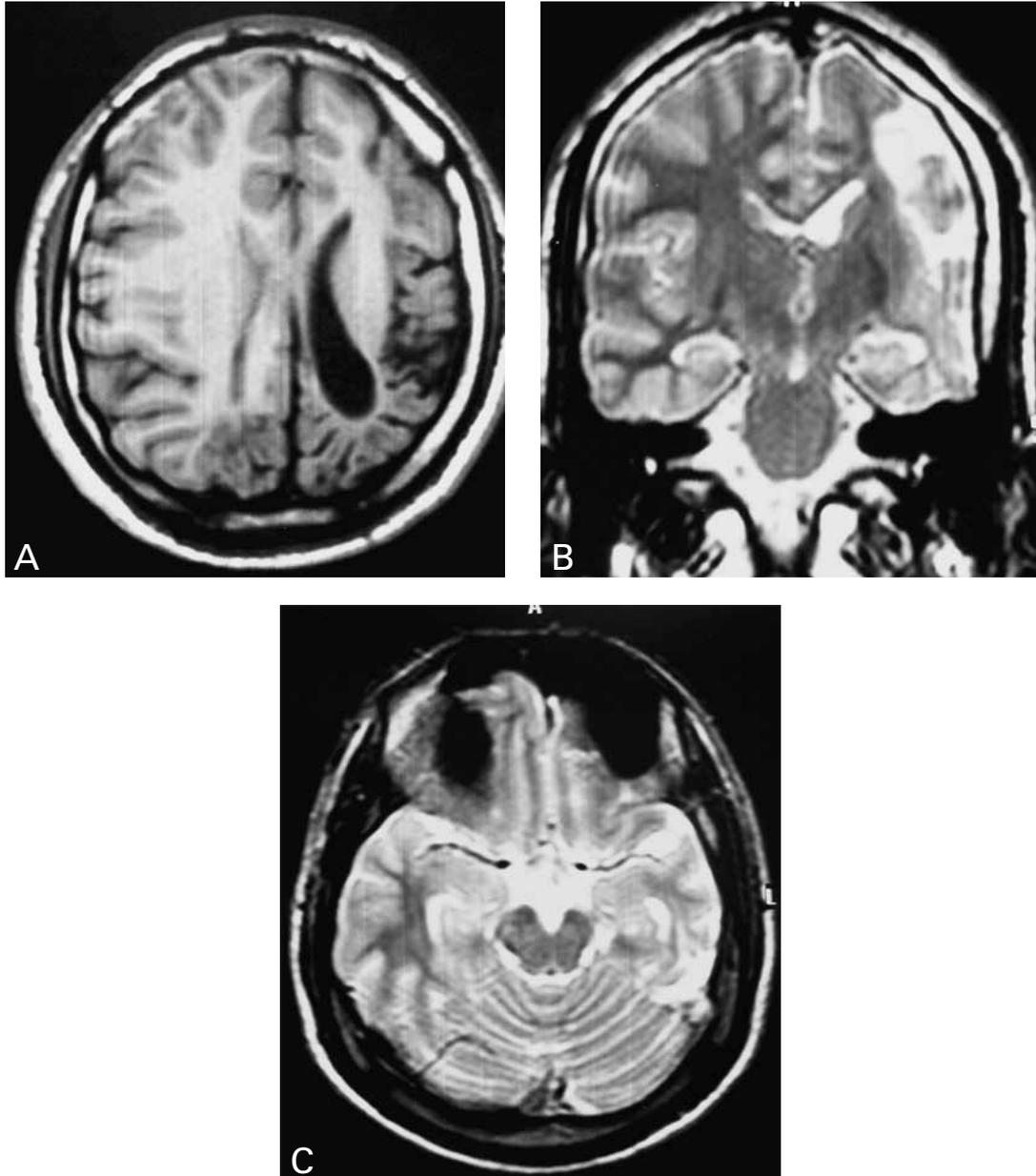


Fig.

1A	1B
1C	

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Work-up

MRI examination of the brain (Fig. 1) shows on the axial T1-weighted image (A) and the coronal T2-weighted image (B) dilated cortical sulci in the left temporal and parietal region. The left ventricle is dilated. Axial T2-weighted image (C) demonstrates diffuse cerebellar atrophy and overpneumatization of paranasal sinuses.

Radiological diagnosis

Based on imaging findings the diagnosis of *Dyke-Davidoff-Masson syndrome* was made.

Discussion

Cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome (DDMS) is a condition characterized by seizures, contralateral hemiplegia or hemiparesis, craniofacial asymmetry and learning difficulties. In 1993 Dyke, Davidoff and Masson described cranial asymmetry on the skull radiographs of nine patients with hemiplegia. Since then this condition was named as the DDMS. Etiological factors can be congenital malformations, vascular occlusion and infection during the prenatal period, birth trauma, anoxia, hypoxia and intracranial hemorrhage during the perinatal period and trauma, infection, prolonged febrile seizure and tumor in the postnatal period. In patients with DDMS due to congenital etiology some compensatory skull changes occur as a result of the adaptation to the atrophy of the brain substance. These compensa-

tory changes include ipsilateral osseous hypertrophy, hyperpneumatization of the ipsilateral paranasal sinuses, enlargement of mastoid cells, elevated temporal bone, hypoplasia of the anterior/middle cranial fossa and shift of the midline structures towards the atrophic hemisphere. Ipsilateral compensatory skull changes are not observed in acquired cases. Three patterns of cranial hemiatrophy have been described on MRI: pattern 1 with diffuse cortical and subcortical atrophy; pattern 2 with diffuse cortical atrophy associated with expanded porencephalic cysts, and pattern 3 with old infarction with necrosis in the middle cerebral arterial territory. Our patient showed the characteristics of pattern 1. In patients with DDMS clinical history is important to determine the etiology of the syndrome. Neuroradiologic workup is necessary to evaluate the extent of the condition.

Bibliography

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