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Case Report

Dyke-Davidoff-Masson syndrome in an 8-year-old child: Report of a case ☆

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ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare entity. Few cases have been described in the literature. It can be symptomatic or asymptomatic. The clinical signs are very varied. Imaging is the key to diagnosis. Calvarial thickening, enlargement sinus, and cerebral hemiatrophy are suggestive signs. It is a cause of cerebral hemiatrophy and epilepsy. We report the clinical and radiological signs of this syndrome through a case of an 8-year-old male child treated for epilepsy. The importance of our article is to report a case diagnosed at an early age (8 years). Most studies report cases diagnosed in adults. MRI revealed pathognomonic signs of Dyke-Davidoff-Masson syndrome.

Case report

The child was an 8-years-old born full term. The parents are not consanguineous with no significant antenatal or perinatal history. He has mental retardation. He has good psycho-motor development. Followed since the age of 6 months for epilepsy disease, there was no history of meningitis, hospital history of epileptic seizures, delayed development of motor skills, and recent left hemicorp heaviness. The clinical examination was normal, and the electroencephalogram was normal too. The patient has never had any imaging. He was referred to radiology for an MRI due to an increase in the frequency of conclusive seizures. The MRI showed enlargement of the subarachnoid spaces of the right convexity, the perisylvian fissure, with discrete enlargement of the choroidal fissure. Signal abnormalities in the right hemispheric cortico-subcortical area, in T2 and FLAIR hypersignal, without reflection on the diffusion sequences and not enhanced after injection of gadolinium, responsible for an attraction of the occipital horn of the homolateral ventricle in relation to an area of gliosis, associated with right hemispheric cortico-subcortical atrophy, homolateral thickening of the cranial vault, and hyperpneumatization of the right frontal sinus (Fig. 1).

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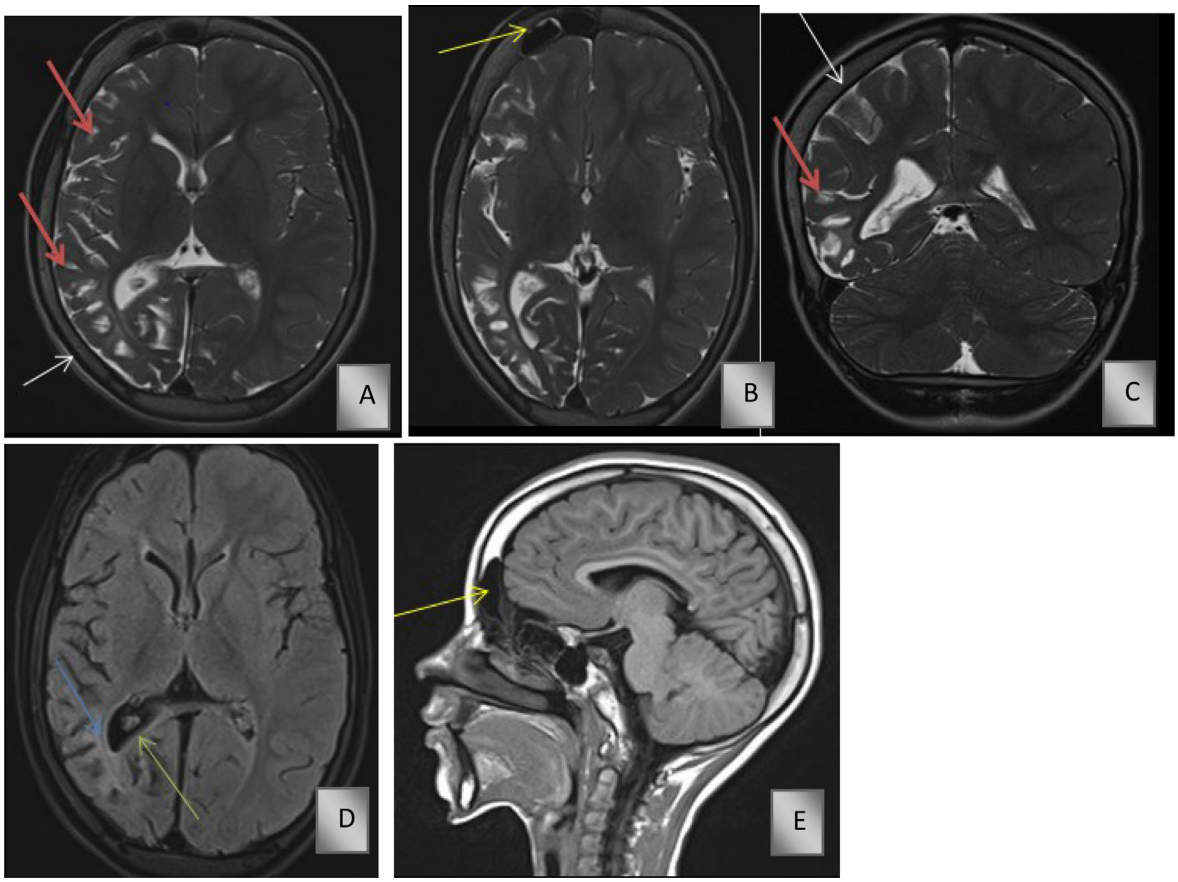

Fig. 1 – Axial (A and B) and coronal (C) T2-weighted images demonstrate: enlargement of the subarachnoid spaces of the right convexity (red arrows), of the sylvian fissure, discrete enlargement of the choroidal fissure. Axial (D) FLAIR: signal anomaly in the right hemispheric cortico-subcortical area (blue arrows) responsible of attraction on the occipital horn of the lateral ventricle (green arrow) in relation to an area of gliosis. Sagittal (E) T1: frontal sinus is hyperpneumatized (yellow arrow) with hypertrophy of cranial vault (white arrow).

Discussion

Dyke-Davidoff-Masson syndrome is a rare entity, described for the first time in 1933. This is a cause of epilepsy and cerebral hemiatrophy. It can affect both adults and children [1]. Diagnosis is made between ages 8 and 75 [2] with a median age of 20 years [14]. It can be congenital or acquired [3]. The pathogenesis is still unclear. During the perinatal period, it may be secondary to a cerebral aggression such as an infection, or vascular damage and coarctation of the midaortic arch [4]. Some hypotheses involve premature closure of the sylvian artery during the perinatal period [5]. The secondary form can occur in the aftermath of an infection, trauma, prolonged febrile seizure hemorrhagic states [7], or cerebral ischemia [2]. The disease can affect both sexes. However, the male sex is more affected. Both hemispheres can be affected predominantly on the left one [6,7]. The development of bone structures and brain parenchyma are harmonious. However, in the case of abnormal development of a part of the brain, it will result in calvaria hypertrophy with hyperpneumatization of the sinuses [8].

Clinical presentation: DDMS represents a spectrum with variable degree of clinical expression. It associates epileptic seizures (often refractory = the most described symptom), can be either focal or generalized [9] mental retardation, hemiplegia, facial asymmetry, cavus foot, cleft palate, atrophy of a hemitronc or of the extremities [5,10]. Coffee spots or ocular lipoderma have been reported, taurodontism and hypoplasia [11]. However, it may be asymtomatic [2]. The severity of the disorder depends on when it occurs during brain maturation. According to Tatlidede et al. [12], some children were still able to lateralize the dorsal visual pathways despite severe symptoms.

The radiological diagnosis is easy in the presence of suggestive signs. On a plan radiograph, we can have calvarial thickening with enlargement of the paranasal sinuses more marked in the frontal sinus because it is the last to be pneumatized. On ultrasound, we can find a hemi-atrophy with “shifted falx” signs [11]. CT scans or, better yet, on magnetic

resonance imaging (MRI) can show unilateral atrophy of the cerebral parenchyma, enlarged sulci and cerebrospinal fluid spaces, enlargement of the homolateral lateral ventricle, and thickening of the cranial vault. In other cases, a compensatory hypertrophy of the contralateral hemisphere may occur [11]. Other signs such as polymicrogyria, peduncular atrophy [5] secondary to Wallerian degeneration may be present thalamic atrophy may also be noted [11,13]. Contralateral cerebellar atrophy may be present in DDMS, but is less frequent than ipsilateral cerebellar atrophy [14]. The presence of gliosis suggests a vascular or infectious cause.

Differential diagnosis can be made with multiple pathologies with similar clinical symptoms:

- Rasmussen encephalitis: Rasmussen encephalitis is a chronic localized encephalitis, characterized by a progressive and unilateral cerebral inflammation of uncertain etiology [15].
- Hemimegaloencephaly: Hamartomatous overgrowth of one part or one hemisphere due to defects in neuronal proliferation, migration and organization; it associates an enlarged cerebral hemisphere and hemicranium with a large homolateral ventricle, SG abnormalities such as pachygvria polymicrogyria, heterotopia, and SB abnormalities [16].
- Sturge-Weber syndrome: Phakomatosisdue to abnormalities in the development of the cortical veins. It is associated with a large spectrum of intracranial manifestations: chronic ischemia with enlargement of alternative drainage veins, and is characterized by a “wine-red” facial capillary malformation +++in the trajectory of the trigeminal nerve [17,18].

Other differential diagnoses include Parry-Romberg syndrome, basal ganglia germinoma, Silver-Russel syndrome, linear nevus syndrome, and Fishman syndrome.

Treatment is based on anticonvulsants in combination with physiotherapy, occupational therapy, and speech therapy. In the event of invalidating seizures, hemispherectomy is recommended [11]. According to the studies, the prognosis of the disease is better when the disease occurs after 2 years [14].

Conclusions

DDMS represents an entity with a very large clinical expression. It is a cause of cerebral hemiatrophy and epilepsy .It may be suspected when clinical signs are suggestive. Or it may be discovered incidentally. MRI is gold standard for best diagnostic tool. The importance of the clinical signs will depend on the level of damage, the period of cerebral maturation, and neuroplasticity.

Author contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

Patient consent

Written informed consent for publication was obtained from patient.

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