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Dyke-Davidoff-Masson Syndrome a Rare Cause of Cerebral Hemiatrophy: A Case Report

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Abstract **Case Report**

Cerebral hemiatrophy or Dyke-Davidoff-Masson syndrome (DDMS) is a rare clinical and radiological entity, characterized by seizures, facial asymmetry, contralateral hemiplegia or hemiparesis and mental retardation. These findings are due to a brain lesion that may occur early in life or in utero. Radiological features include unilateral loss of brain volume associated with compensatory bony alterations in the calvarium, such as thickening, hyperpneumatization of the paranasal sinuses and mastoid cells and elevation of the petrous crest. We report a case of a 13-year-old adolescent who presented with multiple episodes of generalized tonic-clonic seizures. In our case, the clinical history and CT imaging were sufficiently accurate to diagnose DDMS.

Keywords: Dyke-Davidoff-Masson syndrome; Cerebral hemi-atrophy; Imaging.

INTRODUCTION

Dyke-Davidoff-Masson syndrome refers to variable degrees of hypoplasia or atrophy of one cerebral hemisphere with compensatory changes of the calvarium. Dyke, Davidoff and Masson first described the syndrome in 1933 on plain skull radiographs and pneumo-encephalograms in a series of nine patients. The clinical features depend on extent of brain injury and include hemiparesis or hemiplegia, seizures, mental retardation or learning disability, speech or language disorders and facial asymmetry. Imaging studies such as CT and MRI used to make a diagnosis in correlation with clinical features. As it is a rare disorder, it may be misdiagnosed and consequently mismanaged by the majority of physicians [1-3].

CASE PRESENTATION

A 13-year-old child presented with complaints of generalized tonic-clonic epileptic seizures, which started at the age of 11. The frequency of convulsions increased progressively, and even more during the last four months. The child also had a history of speech and language retardation. His family history was unremarkable. On examination, the patient presented with spastic left-sided hemiparesis. Treatment history revealed that the patient had consulted a local physician and was receiving carbamazepine. However, the patient had recently become unresponsive. A cerebral CT scan showed atrophy of the right cerebral hemisphere with enlargement of the cortical sulci in front, causing ipsilateral falciform attraction with a dilated lateral ventricle. Bone window showed thickening of the right hemispheric diploea with enlargement of the homolateral sphenoidal hemisinus and hyper aeration of the right mastoid cells, visible in the temporal scale and small wing of the sphenoid (Figures 1, 2). After correlating the clinical presentation and imaging findings, a final diagnosis of DDMS was made. His treatment regimen consisted of anti-epileptic medications and physiotherapy. The patient was referred to a neurology service for targeted interventions and management.

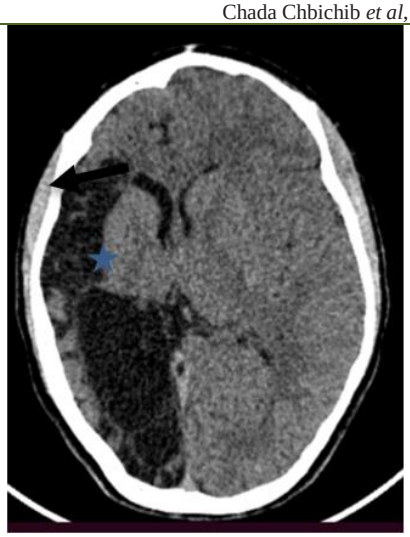

Figure 1: Plain CT head showing atrophy of right cerebral hemisphere (arrow) with dilated ipsilateral lateral ventricle (star), widening of sulci and atrophy of gyri on the right side

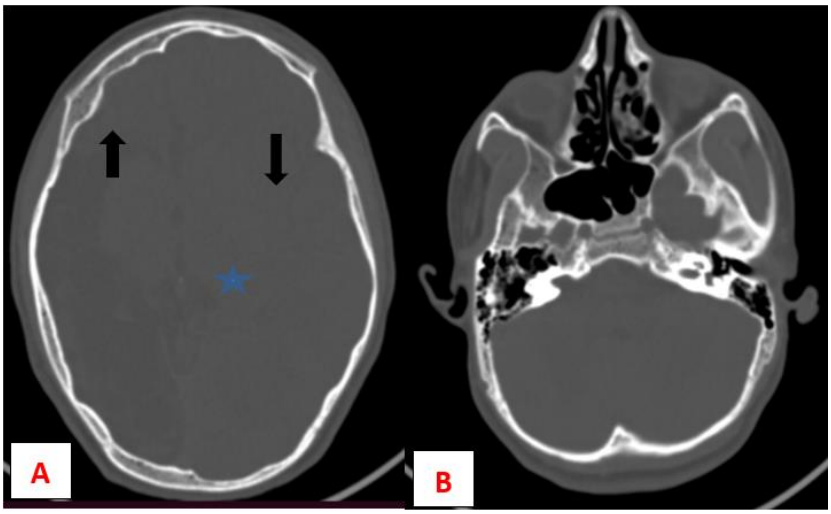

Figure 2: CT head bone window axial images A and B showing thickening of the right hemispheric diploea (arrow up) with enlargement of the homolateral sphenoidal hemisinus (arrow down) and hyperaeration of the right mastoid cells (bleu star)

DISCUSSION

This syndrome was described for the first time in 1933, it covers an imprecise nosological framework with a wide variety of clinical manifestations. The association of epilepsy, usually generalized, facial asymmetry, contralateral motor deficit and mental retardation suggests the diagnosis [5]. Predominantly, there is no established sex predilection or involvement of a specific hemisphere but the involvement of the left side and male gender are more common in the literature [6]. The etiopathogenesis could be either vascular insult during intrauterine life resulting in hypoplasia of a cerebral hemisphere, acquired causes like trauma, infection, vascular abnormalities and intracranial hemorrhage in the perinatal period, or shortly thereafter causing hemi cerebral atrophy [7]. Cerebral atrophy results in turn from a reduction in the formation of brain-derived neurotrophic factors due to these causes [8]. The diagnosis is based on the typical radiological features on computed tomography (CT) and MRI scans which include prominent cortical sulci, encephalomalacia, gliosis, porencephaly, loss of white and gray matter substance, hypoplastic cerebral peduncle, thalamus and internal capsule, ventricular enlargement and midline shift toward the atrophic side. And as a compensatory reaction of the skull, there will be a thickening of the ipsilateral calvaria (diploid space and internal table), hyper-pneumatization of the paranasal sinuses (especially frontal) and mastoid air cells, elevation of the petrous crest, sphenoid wing and orbital roof, reduction in the size of the middle/anterior cranial fossae and displacement of the falx attachment. These radiological features will become more evident over time, as the patient ages [9-11]. This condition should be differentiated from basal ganglia germinoma, Sturge Weber syndrome, Silver-Russel syndrome, linear nevus syndrome, Fishman syndrome and Rasmussen encephalitis. A proper clinical history will provide the correct diagnosis. Treatment should focus on seizure

control with appropriate anticonvulsants. Sometimes, multiple anticonvulsants are used. Hemispherectomy was in dicated in patients with hemiplegia and intractable disabling seizures and is successful in 85% of the cases. In addition to medication, physiotherapy, occupational therapy and speech therapy play an important role in the long-term management of the child [4].

CONCLUSIONS

Because of its rarity, Dyke-Davidoff-Masson syndrome can easily go undiagnosed by the majority of treating clinicians. Knowledge of its features on imaging allows rapid and accurate diagnosis - enabling appropriate management.

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