

CASE REPORT

DYKE-DAVIDOFF-MASSON SYNDROME: A RARE IMAGING CASE

J. B. NDAHIRIWE^{1,*}, E. Cook²

¹Department of Medical Imaging Sciences, Kigali Health Institute

²Education Institute-Fontys University

ABSTRACT

Cerebral hemiatrophy is not frequently encountered in clinical practice of pediatrics. In this case report, we discuss a twelve-year-old male child who presented with seizures since the age of 1 year and left-sided hemiplegia. The CT scan diagnosed the child to have Dyke-Davidoff-Masson Syndrome (DDMS).

Keywords: Cerebral-Hemiatrophy-Syndrome-Seizures.

RESUME

L'hémi-atrophie cérébrale n'est pas rencontrée dans la pratique courante de la pédiatrie. Dans ce cas d'étude, nous discutons un cas d'un garçon âgé de douze ans qui a présenté des convulsions avec une hémiplégie gauche, il était âgé d'un an. La tomodensitométrie (Scanner) a démontré que l'enfant avait le Syndrome de Dyke-Davidoff-Masson (DDMS).

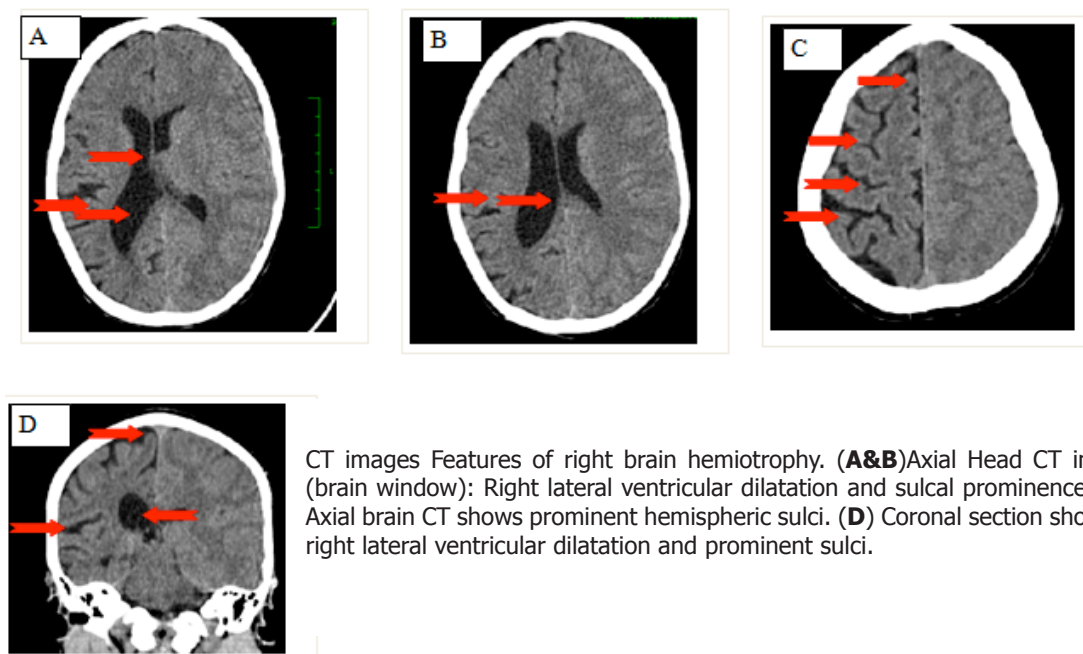
Mots-clés: Cérébral- Hémiatrophie-Syndrome-Convulsions

INTRODUCTION

The Dyke-Davidoff-Masson Syndrome (DDMS) is defined as the atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy) which is secondary to brain insult in fetal or early childhood period. Hemiatrophy is rarely encountered in clinical practice. A 12-year-old boy was referred for CT-scan with left hemiplegia, learning difficulties and seizures for 11 years. There was no clear history of apparent perinatal and antenatal complications.

The boy presented with normal vision and hearing, no face asymmetry was appreciated.

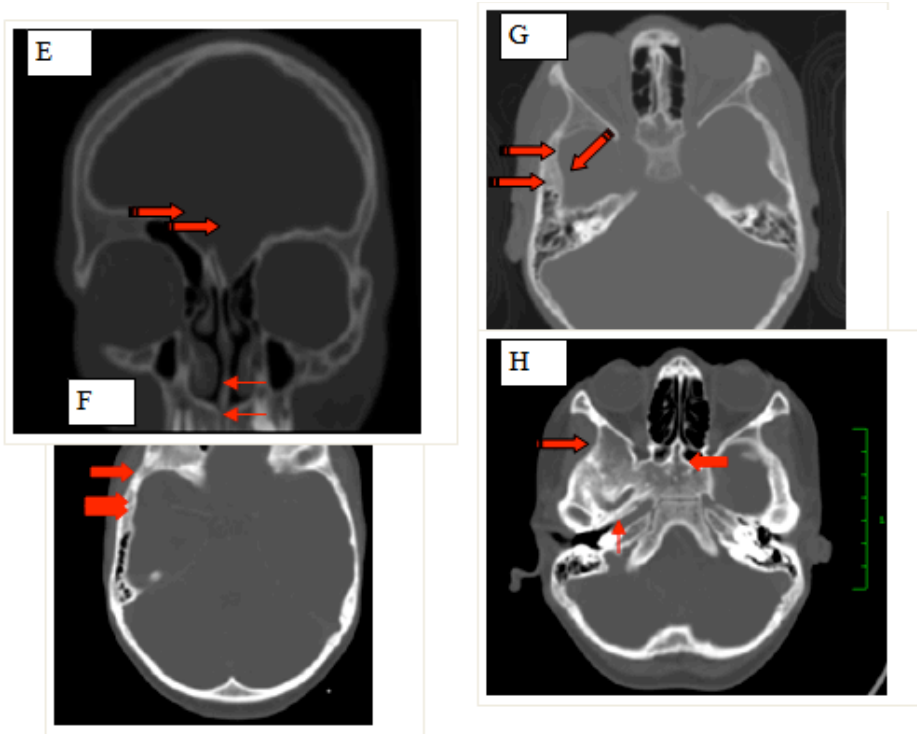
Pre and post contrast CT Scans were performed with 3-6 mm axial slices and Multiplanar Reformatting (MPR) reconstructions (See images A, B, C, D, E, G, F & H).



CT images Features of right brain hemiotrophy. (A&B) Axial Head CT image (brain window): Right lateral ventricular dilatation and sulcal prominence. (C) Axial brain CT shows prominent hemispheric sulci. (D) Coronal section showing right lateral ventricular dilatation and prominent sulci.

*Correspondence to:

Jean Baptiste NDAHIRIWE
Department of Medical Imaging Sciences, Kigali Health Institute



CT images **E** (coronal section) & **F** (axial section): Right frontal sinus widening with extension the ethmoid sinus

CT images **G & H** (axial section): Right calvarial thickening (including right squamous portion of the temporal bone). Widened mastoid air cells Elevation of the greater wing of sphenoid and petrous ridge on the right side

DISCUSSION

The CT brain shows the right cerebral hemispheric atrophy with ex vacuo dilatation of the right lateral ventricle, thickening of right frontal calvarium with the squamous portion of the temporal bone and widening (enlargement) of the right frontal sinus and mastoid air cells. There is midline shift to the right side and right hemisphere sulcal prominence and ipsilateral sylvian fissure. There is elevation of the greater wing of sphenoid and right petrous ridge. These findings are consistent with the diagnosis of Dyke-Davidoff-Masson syndrome (DDMS). There was no need to recommend a Magnetic resonance scanning.

Dyke-Davidoff-Masson syndrome was described as skull radiographic and pneumoencephalographic changes in their series of 9 patients whose clinical characteristics included hemiparesis, seizures, facial-asymmetry, and mental retardation in 1933 by Dyke-Davidoff and Masson [1].

It has been reported that DDMS is caused by cerebral insult that may occur in utero when the maturation of calvarium has not been completed, or during early life due to brain damage (usually traumatic) [1, 2].

Prenatal causes include congenital abnormalities, cerebral infarction, vascular malformations and infections. Perinatal causes include birth trauma, hypoxia and intracranial hemorrhage. Finally, cerebral hemi-atrophy can develop secondary to cerebral trauma, tumors, infections and prolonged febrile seizures after birth [2]. Generally, the etiological factor for Dyke-Davidoff-Masson syndrome

has been postulated as trauma, inflammation or vascular malformations and occlusions. When the insult occurs in-utero, it could be due to gestational vascular occlusion, primarily involving the middle cerebral vascular (MCA) territory.

According to Sharmal et al decreased carotid artery blood flow due to coarctation of aorta can also cause cerebral hemiatrophy [3]. Garg et al reported a possible etiological relation of cerebral hemiatrophy with febrile seizures. In their three cases of cerebral hemi atrophy, Sener and colleagues, reported the middle cerebral artery stroke to be the cause [4, 5].

DDMS is seldom seen in clinical practice. When this develops early in life (during the first two years), certain cranial changes like ipsilateral hypertrophy of the skull and enlargement of sinuses occur, the elevations of the greater wing of sphenoid and the petrous ridge on the affected side. The compensatory cranial changes occur to take up the relative vacuum created by the atrophied or hypoplastic cerebral hemisphere [6].

The classical clinical presentation includes seizures, facial asymmetry, contralateral hemiplegia or hemiparesis and mental retardation. However, according to Setty et. al., mental retardation is not always present and seizures may appear months or years after the onset of hemi paresis. The clinical findings may be of variable degree according to the extent of the brain injury. Imaging studies show unilateral loss of volume of brain and calvarial changes, findings

of cerebral atrophy, ex-vacuo ventricular dilatation and enlargement of sulci [3, 7].

There are two types of DDMS: congenital or infantile and acquired. In congenital hemiatrophy, when the insult occurs in-utero, there is shift of midline structures towards the side of the disease and the sulcal prominence replacing the gliotic tissue is absent [8]. This feature differentiates it from cerebral hemiatrophy which occurs in early life. The atrophied cerebral hemisphere will have prominent sulcal spaces if the insult occurs after birth or after end of sulcation [11].

A comprehensive clinical history and CT or MRI provides the right diagnosis. In addition to CT findings described above, MRI demonstrates the gray white matter loss with hyperintensities on T2 weighted images (diffuse cortical and subcortical atrophy) and asymmetry of the basal ganglia [8]. MRI has the ability to bring to light changes in the cerebral hemispheres as well as highlighting bony structures, thus differentiating between congenital and acquired types of DDMS [6].

DDMS should be differentiated from Basal cell germinoma, Sturge Weber syndrome, Linear-Nevus syndrome, Fishman syndrome, Silver-Russell syndrome and Rasmussen encephalitis [9].

The treatment is symptomatic, and should target convulsion, hemiplegia, hemiparesis and learning difficulties. Prognosis is better if hemiparesis occurs after the age of 2 years and in absence of prolonged or recurrent seizures. Children with intractable disabling and hemiplegia are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases [10].

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