SUBMITTED 21 APR 22
REVISION REQ. 21 JUL 22; REVISION RECD. 2 AUG 22
ACCEPTED 24 AUG 22
ONLINE-FIRST: September 2022
DOI: https://doi.org/10.18295/squmj.9.2022.055

Imaging Features of Dyke-Davidoff-Masson Syndrome

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Introduction

A 42-year-old man presented to the adult epilepsy clinic in 2019 at Sultan Qaboos University Hospital, Muscat, Oman, with right motor seizures with occasional generalization and loss of consciousness, right-side hemiparesis, and low intellectual capacity since early childhood. The patient had perinatal hypoxic ischemic injury due to a difficult delivery, which resulted in spastic cerebral palsy. His developmental history revealed delayed milestones and learning difficulties at school. His seizure episodes have increased in frequency over the years, requiring treatment with multiple antiepileptics. Despite being on four antiepileptic drugs, the patient continued to experience very frequent seizures. On examination, he had slurred speech and right facial deviation. He was able to walk without support, with a circumduction gait. There was right-sided spastic hemiparesis with brisk tendon reflexes and an extensor plantar response.

An electroencephalogram (EEG) was done and showed left hemispheric cerebral dysfunction with left hemispheric onset focal motor seizures. Computed tomography (CT) and magnetic resonance imaging (MRI) of the brain were done and revealed atrophy of the left cerebral hemisphere with ipsilateral compensatory thickening of skull bones and hyperpneumatization of the left frontal and sphenoidal sinuses. The diagnosis of Dyke-Davidoff-Masson (DDM) syndrome was made based on clinical and radiological findings. The patient was managed with antiepileptic medications, muscle relaxants, and physiotherapy. Due to his intractable seizures,

he was offered surgical intervention at an outside facility. We present a case of DDM syndrome with characteristic clinical and radiological findings. It is a rare entity with few reported cases in literature. To best of our knowledge, this is the first case to be reported in our institution.

Informed patient consent for publication was obtained.

Comment

DDM syndrome is a rare condition that was described for the first time in 1933 by Dyke, Davidoff, and Masson.[1,2] Those researchers described the plain skull radiograph features of nine patients who presented with seizures, hemiparesis, facial asymmetry, and mental retardation.[1]

There are two types of DDM syndrome: congenital and acquired.[2] The congenital or infantile type presents early in infancy, secondary to previous intrauterine brain insults like vascular occlusion or anomaly of the middle cerebral artery mainly.[2,3] The acquired type occurs later in childhood secondary to variable causes affecting brain perfusion like infection, prolonged febrile seizure, trauma, hemorrhage, or ischemia.[2] Our patient presented with hemiplegia and seizures since childhood, which is likely due to the hypoxic ischemic injury that he sustained during the perinatal period. The classical imaging features of this condition are unilateral cerebral atrophy with ipsilateral calvarial thickening and hyperpneumatization of the ipsilateral frontal sinus.[1,3] Other reported findings are ipsilateral falcine displacement, elevation of the petrous ridge and wing of sphenoid bone, atrophy of ipsilateral basal ganglia and brainstem and contralateral cerebellum, and hyperpneumatization of ipsilateral mastoid air cells.[2]

Our patient has the classic clinical and imaging features described in the literature for DDM syndrome. He had CT [Figure 1] and MRI [Figure 2] of the brain, which revealed unilateral left-sided cerebral atrophy with ipsilateral compensatory hypertrophic thickening of skull bones. There was also hyperpneumatization of the left frontal and sphenoidal sinuses and the left mastoid process. Atrophy of the ipsilateral thalamus, cerebral peduncle, and falttending of the left anterior surface of the pons and medulla in keeping with Wallerian degeneration, and contralateral cerebellar atrophy were present. On time-of-flight MR angiogram [Figure 3], the left middle cerebral artery and its branches were smaller compared to the right side. It is

noteworthy to mention that the imaging findings of cerebral hemiatrophy, features of Wallerian degeneration, and cossed cerebellar diaschisis due to impaired neuronal connections can be seen after various major cerebral inulsts, commonly infarcts, at any time in life. The presence of the findings varies depending on the severity of the insults. The presence of skull and sinus hypertrophy is seen, however, in insults that happen early in life, as classically described in DDM syndrome.

The most common differential diagnosis for this syndrome is chronic Rasmussen encephalitis, Sturge-Weber syndrome, basal ganglia germinoma, and Fishman syndrome.[1,2] However, differentiation between them can be made by clinical and radiological findings. For example, chronic Rasmussen encephalitis is a rare progressive inflammatory disease affecting children who usually present with seizures and cognitive impairment. Imaging findings show unilateral cerebral atrophy without associated skull changes.[1] Patients with Sturge-Weber syndrome usually present with seizures, mental retardation, and a typical port wine stain on the face in the distribution of the ophthalmic division of the trigeminal nerve. On imaging, there will be unilateral cerebral atrophy with increased angiomatosis and sometimes ipsilateral cortical tram track calcifications.[2,3] DDM syndrome has a wide spectrum of presentation, ranging from mild symptoms to severely disabling symptoms.

Refractory seizures remain the most challenging complaint.[4,5] Patients with refractory seizures may benefit from surgical interventions like functional hemispherectomy, with a reported success rate of 85% in selected cases.[1,4] However, it is highly associated with long term adverse effects like obstructive hydrocephalus and chronic subdural hygromas.[2,4] Hemispherotomy is another surgical option that is also effective and associated with fewer complications. Rehabilitation, physiotherapy, speech therapy, and occupational therapy are important to improve the quality of life.[4,5] Our patient was managed with antiepileptic medications, muscle relaxants, and physiotherapy. Due to his intractable seizures, he was offered surgical intervention at an outside facility.

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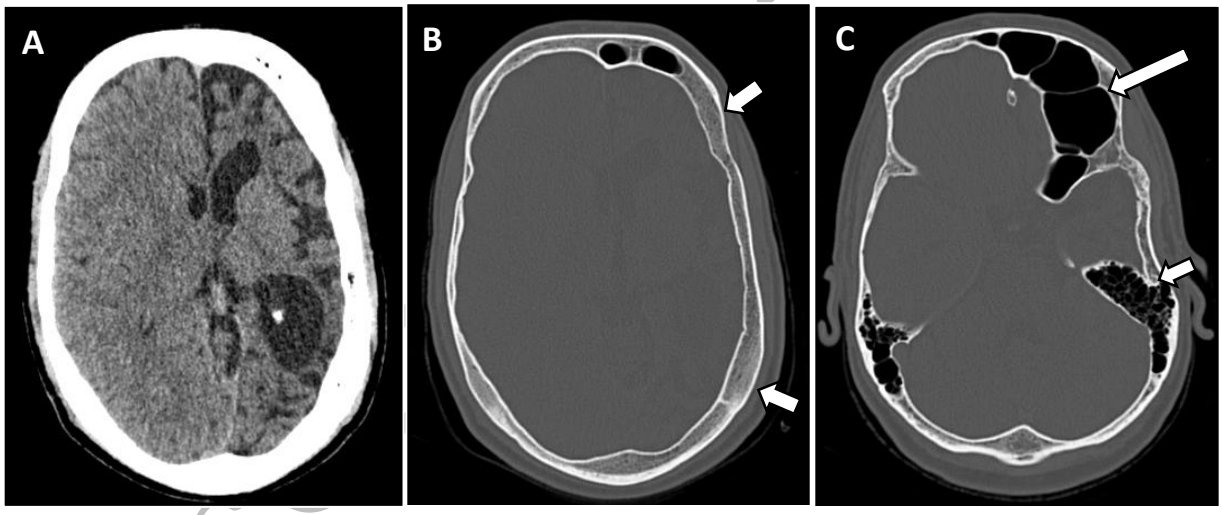

Figure 1: Axial unenhanced CT of the brain with brain and bone windows shows **(A)** severe left-side cerebral atrophy with ex-vacuo dilatation of the ipsilateral lateral ventricle, **(B)** left-side compensatory calvarial hypertrophy (arrows), **(C)** hyperpneumatization and enlargement of the left frontal sinus (long arrow) and left mastoid air cells (short arrow).

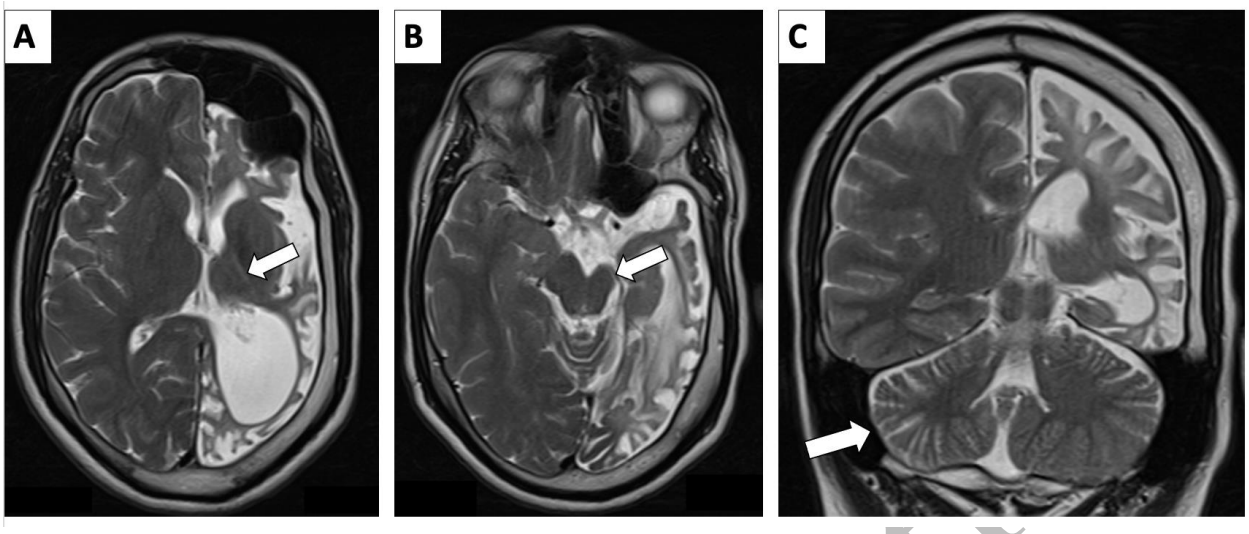

Figure 2: Axial (A,B) and coronal (C) T2 weighted images demonstrate severe atrophy of the left cerebral hemisphere with (A) left thalamic atrophy (arrow), (B) left cerebral peduncle atrophy (arrow) and (C) contralateral cerebellar atrophy (arrow).

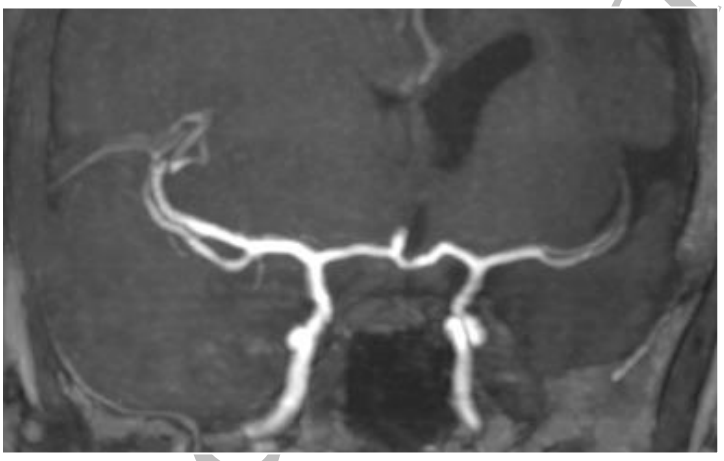

Figure 3: Coronal time-of-flight MR angiogram: The left main cerebral artery and its branches are smaller than the right side. (arrow).