DYKE-DAVIDOFF-MASSON SYNDROME SECONDARY TO VASCULAR COMPROMISE: A RARE CAUSE OF CEREBRAL HEMIATROPHY

Ashok Kumar, Bushra Rehan, Imtiaz Ali Panhwar, Kanchan

Department of Radiology, Liaquat National Hospital, Karachi, Pakistan.

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ABSTRACT

Dyke-Davidoff-Masson syndrome is a rare neurological condition resulting from brain insult during intrauterine life or in early childhood. Clinical presentation includes seizures, mental retardation, contralateral hemiparesis, and facial asymmetry.[1] Crossectional radiology may show unilateral cerebral hemiatrophy/hypoplasia, ipsilateral ventricular enlargement, ipsilateral hyperpneumatization of the sinuses, and compensatory calvarial thickening.[1] Being a rare case according to literature review,[1] it may be easily missed clinically and radiologically. Here we report a case of an 8 year old male patient who preseted with right sided body weakness, generalized tonic clonic fits and mental retardation. MRI of the patient showed atrophy of left cerebral hemisphere along with dialatation of left lateral ventricle, enlargement of left frontal sinus and compensatory hypertrophy of left side of the clavarium, Wallerian degeneration of left hemi mid brain. The findings were suggestive of Dyke-Davidoff-Masson syndrome and most likely cause in this patient was vascular compromise.

Key words: Cerebral hemiatrophy, Dyke Davidoff Masson syndrome, seizures, hemiparesis.

Introduction

The Dyke-Davidoff-Masson syndrome (DDMS) was introduced for the first time in 1933 by Dyke, Davidoff, and Masson who reported a case series of 9 patients with skull radiographic and pneumatoencephalographic changes having clinical features of hemiparesis, facial asymmetry, seizures, and mental retardation.[2] DDMS is rarely seen in clinical practice. It is caused by brain insult either prenatally or during early childhood.[3] The clinical findings may include seizures, mental retardation, contralateral hemiparesis, and facial asymmetry.[1] Here we present a case of an 8 year old male patient who preseted with right sided body weakness, generalized tonic clonic fits and mental retardation and characteristic radiologic findings were suggestive of Dyke Davidoff Masson syndrome (DDMS) and most likely cause in this patient was vascular compromise.

Correspondence : Dr. Ashok Kumar
Department of Radiology,
Liaquat National Hospital,
Karachi, Pakistan.
Email: ashok.jeythwani@gmail.com

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Case Report

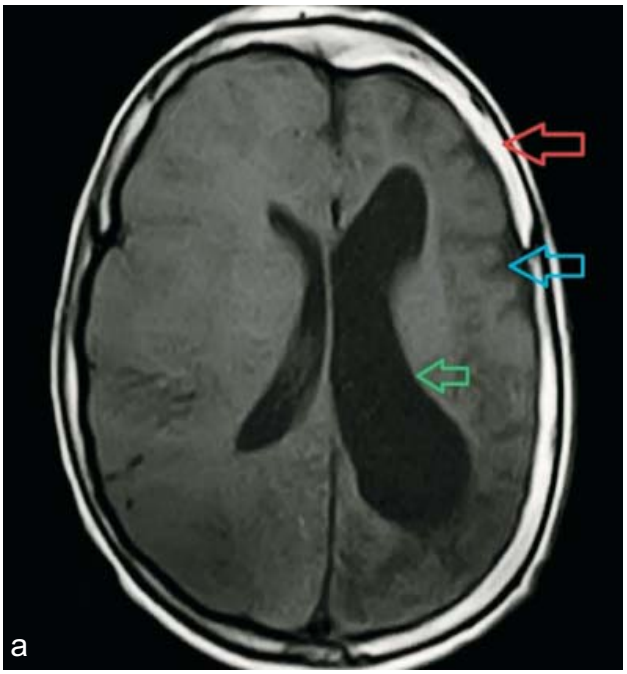
An 8 year old male patient with previous history of perinatal hypoxia presented with right sided body weakness, generalized tonic clonic seizures and mental retardation from 5 years.

On general appearance, the patient was active but was looking mentally retarded. Neurological examination performed by pediatric neurologist revealed 4/5 power in the right upper and lower limb with brisk reflexes. His routine labs were within normal limits.

MRI of the patient showed atrophy of left cerebral hemisphere along with dialatation of left lateral ventricle, enlargement of left frontal sinus and compensatory hypertrophy of left side of the calvarium, Wallerian degeneration of left hemi mid brain. His MRA revealed tapering of M2 segment of left middle cerebral artery with significantly reduced peripheral cortical branches which also show mild to moderate segmental narrowing. Moderate segmental narrowing

was also seen in left posterior cerebral artery with its gradual tapering. Rest of the anterior and posterior circulation appeared to be normal.

On the basis of clinical and radiological findings diagnosis of Dyke-Davidoff-Masson syndrome was made which was likely secondary to vascular compromise as shown by MRA findings.



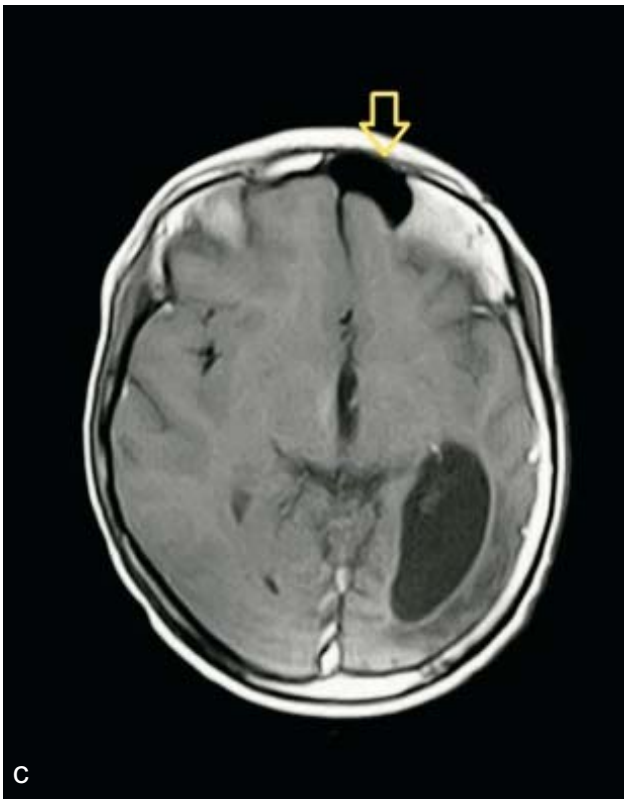
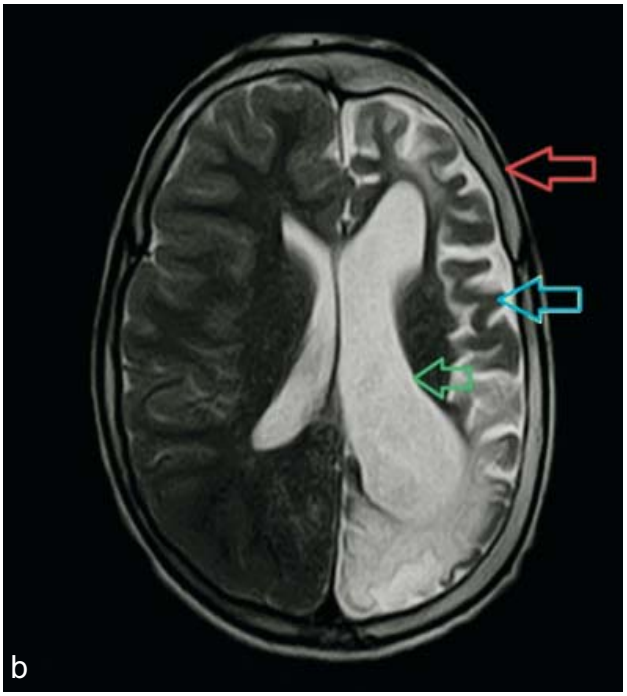


Figure 1: T1 **(A)** and T2W **(B)** images show atrophy of left cereberal hemisphere (blue arrow), dilatation of left lateral ventricle (green arrow) and calvarial hypertrophy (red arrow). T1 images **(C)** shows enlargement of left frontal sinus (yellow arrow).



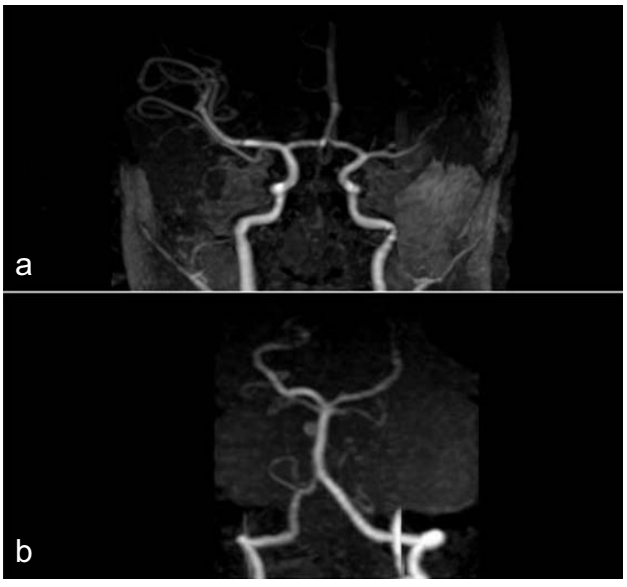


Figure 2: Shows tapering of M2 segment of left middle cerebral artery with significantly reduced peripheral cortical branches **(A)** and moderate segmental narrowing in left posterior cerebral artery with its gradual tapering **(B)**.

Discussion

Dyke, Davidoff, and Masson for the first time in 1933 reported a case series of 9 patients with skull radiographic and pneumatoencephalographic changes who were having clinical features of hemiparesis, facial asymmetry, seizures, and mental retardation.[2] DDMS occurs most commonly in pediatric population but can also rarely be discovered in adult population as well.[1]

DDMS is caused by brain insult that may occur prenatally when the maturation of calvarium has not been completed, or during early life due to brain damage from various causes before the age of 2 years. Prenatal causes may include vascular malformations, intracranial infections, cerebral infarcts and vascular occlusion, primarily involving the middle cerebral vascular territory. peri- and post-natal causes include birth trauma, hypoxia, intracranial hemorrhage, tumors, infections, and prolonged febrile seizures.[3] Perinatal hypoxia is very common in developing countries like Pakistan, due to poor obstetric care.[4] our patient also had previous history of perinatal hypoxia.

The clinical findings in DDMS may include refractory seizures, facial asymmetry, contralateral hemiplegia or hemiparesis, mental retardation, and rarely, patients can also present with sensory symptoms and psychiatric disorders such as schizophrenia.[3]

Crossectional imaging i.e. CT and MRI are very frequently needed for correct diagnosis and early management. Imaging features for DDMS include unilateral cerebral hemiatrophy/hypoplasia, ipsilateral ventricular enlargement, ipsilateral hyperpneumatization of the sinuses, and compensatory calvarial thickening.[1] Any gender can be affected and any cerebral hemisphere can be involved but literature review suggest that it is more frequent in males and usually involve left cerebral hemisphere.[5] Our case is also a male with left cerebral involvement.

Important differential diagnosis includes Sturge-Weber Syndrome, basal cell germinoma, Fishman syndrome, Silver-Russell syndrome, linear nevus syndrome.[3] Rasmussen syndrome is a rare inflammatory disease of children. Seizure and mental retardation are its common clinical features. Cerebral hemiatrophy is common radiological finding but usually there is no calvarial hypertrophy which is a common finding in DDMS. Sturge-Weber Syndrome presents with seizures, mental retardation and a port-wine stain on the face. Radiological findings include cerebral hemiatrophy, calvarial thickening, enlarged sinuses which are similar findings as in DDMS. While in SWS there are also gyriform calcifications, enhancing pial angiomas, enlarged choroid plexus which are not seen in DDMS.

Conclusion

Being a rare cause of cerebral hemiatrophy Dyke-Davidoff-Masson syndrome may easily be missed by the majority of clinicians and radiologists. Adequate knowledge of its clinical and radiological presentation and risk factors may lead to prompt diagnosis and management.

Conflict to declare: None to declare

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