



CASE OF DYKE DAVIDOFF MASSON SYNDROME

Medical Education

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ABSTRACT

Dyke-Davidoff-Masson syndrome is a rare entity characterized by hemi cerebral atrophy/hypoplasia secondary to brain insult in fetal or early childhood period along with ipsilateral compensatory osseous hypertrophy and contralateral hemiparesis. We present two cases of this uncommon condition and discuss its imaging features, differential diagnosis, treatment options and prognosis.

KEYWORDS

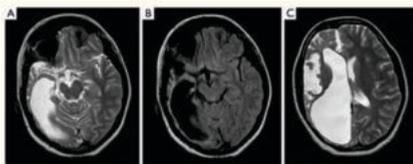
Hemiatrophy, Refractory Seizures, Magnetic Resonance Imaging (MRI), Dyke Davidoff Masson Syndrome, Computed Tomography.

INTRODUCTION

Dyke-Davidoff-Masson syndrome refers to variable degrees of hypoplasia or atrophy of one cerebral hemisphere with compensatory changes of the calvarium. The etiopathogenesis could be either vascular insult during intrauterine life resulting in hypoplasia of a cerebral hemisphere or acquired causes like trauma, infection, vascular abnormalities and intracranial hemorrhage in the perinatal period or shortly thereafter causing hemi cerebral atrophy. Cerebral atrophy in turn results due to reduction in the formation of brain derived neurotrophic factors by these causes.

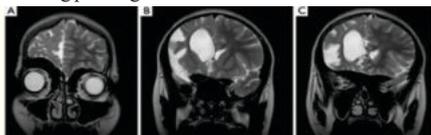
The syndrome was first described by Dyke, Davidoff and Masson in 1933 on plain skull radiographs and pneumo-encephalograms in a series of nine patients. The clinical features depend on extent of brain injury and include hemiparesis or hemiplegia, seizures, mental retardation or learning disability, speech or language disorders and facial asymmetry. Rarely patients can also present with sensory symptoms and psychiatric disorders like schizophrenia. Nevertheless, seizures may appear months to years after the onset of hemiparesis and mental retardation is not seen in all cases.

The diagnosis is based on the typical radiological features on computed tomography (CT) and MRI scans which include cerebral hemiatrophy with dilated ipsilateral lateral ventricle. Also there is thickening of calvarium with enlargement of frontal, ethmoid and sphenoid sinuses and elevation of greater wing of sphenoid and petrous ridge. The osseous abnormalities can also be depicted on plain skull films.



(A-C) Axial T2 weighted and (B) FLAIR sequence showing (A&B) right hemi cerebral atrophy with enlarged right frontal sinus.

The differential diagnoses are chronic Rasmussen encephalitis (chronic, progressive inflammation of brain of uncertain etiology) and Sturge weber syndrome. However, Rasmussen encephalitis doesn't show calvarial changes and Sturge-weber syndrome additionally shows enhancing pial angiomas and cortical calcifications .



Coronal T2 weighted images showing hemi atrophy of right cerebral hemisphere with (A) enlarged right frontal sinus, (B) sphenoid sinus and (C) elevated sphenoid ridge on the right.

Management consists of control of seizures with appropriate anticonvulsants, as most patients with this disorder present with refractory seizures. Additionally, domiciliary physiotherapy, occupational and speech therapy have a crucial role. Hemispherectomy is indicated in patients with hemiplegia and intractable disabling seizures and is successful in 85% of the cases. Prognosis is poor in cases of prolonged or recurrent seizures and if hemiparesis occurs before two years of age. Hence, it is indeed very important for neurologists and radiologists to be familiar with this condition for its early diagnosis and treatment.

Case Report

A 43-year-old female patient presented to the casualty with history of right sided upper limb weakness, vomiting, decrease response to commands and altered mental sensorium.

History of left upper and lower limb weakness since childhood.

Past History:

- Known case of seizure disorder since 42 years and is non-compliant to medication.
- Known case of poliomyelitis.

On Examination:

- BP- 110/70 mm Hg
- PR- 76 bpm
- RR- 14/ min
- GCS- E4V4M5

Radiological Workup Included:

- MRI Brain Screening
- MR Brain Angio + Veno
- CT Brain.

Mri Brain Screening

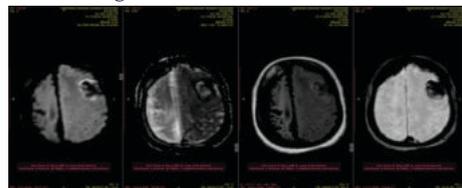


Figure 1

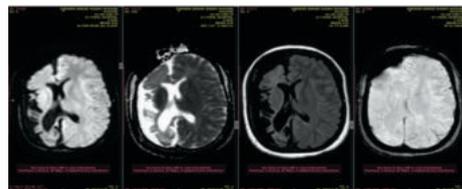


Figure 2

- An area of altered signal intensity which shows significant blooming on gradient images and appears hyperintense on FLAIR with peripheral hyperintense rim is noted involving the left frontal lobe ----Most likely suggestive of intraparenchymal hemorrhage. Few areas of adjacent sulcal blooming are also noted which may suggest minimal subarachnoid hemorrhage. Areas of blooming are also noted involving superior sagittal and right transverse sinus. Advice MRI contrast venogram to rule out superficial venous sinus thrombosis with hemorrhagic venous infarct. (Figure 1)
- There is significant atrophy of the right cerebral hemisphere with associated prominence of the right cortical sulci and right ventricular system as compared to the left resulting in asymmetry of the cerebral hemispheres. There is associated calvarial thickening on the affected side. There is also ipsilateral enlargement of the right frontal sinus----findings suggestive of right sided cerebral hemi atrophy –Dyke Davidoff Masson. (Figure 2)
- Mild atrophy of the right cerebral peduncle and the right ventral hemi mid brain is noted most likely suggestive of wallerian degeneration.

MR BRAIN ANGIO + VENO



MR Venograms were obtained for craniocervical vessels using 2D & 3D TOF sequences.

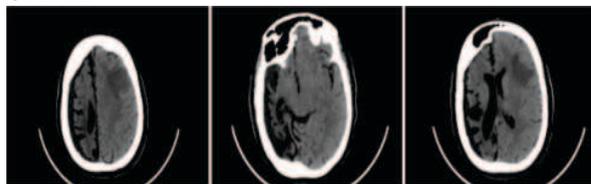
- Non enhancing filling defects which show blooming on gradient images are seen involving entire superior sagittal sinus, entire right transverse, right sigmoid sinuses and jugular bulb suggestive of venous sinus thrombosis. ADVICE: Contrast study for confirmation.
- The right internal cerebral vein is narrowed in caliber as compared to the left
- Few small eccentric areas of blooming are seen in left transverse and left sigmoid sinuses----? arachnoid granulation.
- The vein of Galen is normal.

MR Angiograms were obtained for craniocervical vessels using 2D & 3D TOF sequences.

Suboptimal scan due to motion artifacts limit the evaluation of cervical vessels.

- There is short segment narrowing of A1 segment of left anterior cerebral artery for a length of 5.5 mm causing 80-90 % luminal compromise.
- The right anterior cerebral artery appears narrowed in caliber as compared to the left
- There is significant narrowing of the M3, M4 segments of the right middle cerebral artery, with attenuation of caliber of M1, M2 segments of right middle cerebral artery as compared to the left.

CT Brain



- A hypodense area is noted involving the left frontal region. It is causing mass effect in the form of effacement of the overlying cortical sulci. Findings are most likely suggestive of resolving intraparenchymal hemorrhage. (Figure A and C)
- There is significant atrophy of the right cerebral hemisphere with associated prominence of the right cortical sulci and right ventricular system as compared to the left resulting in asymmetry of the cerebral hemispheres. There is associated calvarial thickening on the affected side. There is also ipsilateral

enlargement of the right frontal sinus----findings suggestive of right sided cerebral hemi atrophy –Dyke Davidoff masson syndrome. (Figure B)

- Mild atrophy of the right cerebral peduncle and the right ventral hemi mid brain is noted most likely suggestive of wallerian degeneration.

CONCLUSION

Based on the abovementioned imaging features, the possible radiological diagnosis was Dyke Davidoff Masson Syndrome

Other Radiological Differentials Include:

1. Chronic Rasmussen encephalitis (chronic, progressive inflammation of brain of uncertain etiology)
2. Sturge Weber Syndrome.

However, Rasmussen encephalitis does not show calvarial changes and Sturge-Weber syndrome additionally shows enhancing pial angiomas and cortical calcifications.

DISCUSSION

There was presentation of right sided upper limb weakness, vomiting, decrease response to commands and altered mental sensorium with history of left upper and lower limb weakness since childhood.

A positive past history of seizure disorder non-compliant to medication and poliomyelitis.

The GCS at presentation was E4V4M5. MRI Brain Screening was suggestive of Intraparenchymal Hemorrhage with Dyke Davidoff Masson syndrome vs Rasmussens encephalitis. MRI Brain Angio+Veno was done which was suggestive of Central Venous Sinus Thrombosis. CT Brain revealed a resolving intraparenchymal hemorrhage with Dyke Davidoff Masson syndrome.

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