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Dyke-Davidoff Mason Syndrome-A rare case report

KEYWORDS: Dyke-Davidoff-Masson Syndrome, hemiatrophy of brain.

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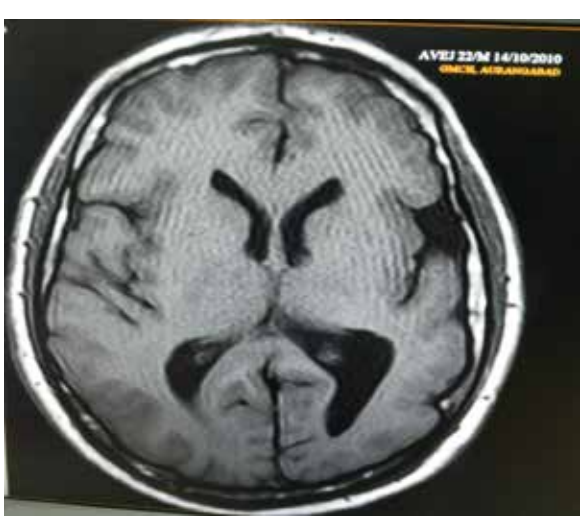
ABSTRACT *A 22 year old patient presented with history of altered sensorium since 3 days. Patient was referred with CSF examination done in private hospital. On admission patient was treated with anticonvulsants. Patient improved. NCCT and MRI brain was done which revealed hemiatrophy of brain. After ruling out other differentials of hemiatrophy, a diagnosis of Dyke Davidoff Mason syndrome was entertained.*

Introduction:
Dyke-Davidoff-Masson syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy) which is secondary to brain insult in fetal or early childhood period.[1] Hemiatrophy is not frequently encountered in clinical practice. We present here a case of a 22 year old man who presented with altered sensorium and on NCCT and MRI brain was diagnosed to have DDMS.

Case history:
A 22 year old patient had presented with history of altered sensorium since the last 3 days. Patient was evaluated and examined. Patient had bilateral plantars extensor on admission. Patient was presumed to be in post-ictal phase. Patient was treated with anti-convulsants. Patient repeatedly had deterioration in consciousness and was treated as non-convulsive status. Patient responded to combination anti-convulsant therapy. Patient was evaluated with NCCT and MRI brain which revealed holohemispheric atrophy on the left side and dilated ipsilateral ventricle and increased diploic spaces. Thus, concluding this as DDMS.



T1 weighted and flair MRI Brain shows holohemispheric atrophy on the left side and dilated ipsilateral ventricle and increased diploic spaces.

Discussion:
In 1933, Dyke,Davidoff and Masson described the plain skull radiographic and pneumatoencephalographic changes in series of nine patients characterised clinically by hemiparesis, seizures, facial-asymmetry, and mental retardation.[1] The plain skull radiographic changes included thickening of calvarium and dilatation of ipsilateral frontal and ethmoid sinuses. Also, there was elevation of great wing of sphenoid and petrous ridge and upward lifting of planum-sphenoidale after brain damage was sustained.[6] A vascular cause of cerebral hemiatrophy (hypoplasia), first proposed in 1860 was confirmed in later studies.[7]

It was proposed that a vascular anatomy occurring in very early gestation(five or six weeks) may result in a major defect in brain development whereas those occurring later may produce more localized lesions.[8] It was reported that decrease in carotid artery flow due to coarctation of aorta can also cause cerebral hemiatrophy. Crossed cerebellar atrophy is usually associated with long standing, extensive

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and unilateral cerebral lesions with onset during infancy or early childhood. The manifestations of DDMS may be so subtle as to be overlooked on plain radiographs; however, CT is the diagnostic modality of choice. Other differential diagnosis to be considered in a patient of cerebral hemiatrophy are Sturge-Weber syndrome, some brain tumours, Silver syndrome as well as conditions that are associated with unilateral megalencephaly like linear-nevus syndrome. A proper clinical history and CT findings provide the correct diagnosis.

In 1939, Alpers and Dear defined two types of cerebral hemiatrophy.[2] In the primary (congenital) type, the entire cerebral hemisphere is characteristically hypoplastic. The secondary type results from a cerebrovascular lesion, inflammatory process or cranial trauma. A clinical triad of hemiplegia, seizures and mental retardation was defined. However mental retardation was not always present and seizures may appear months or years after the onset of hemiparesis.[3]

According to Hageman et al, since primary cerebral atrophy is usually a lack of cerebral development rather than atrophy the terms cerebral hemi-hypoplasia or unilateral cerebral hypoplasia would be more appropriate.[4]

The brain reaches half of its adult size during the first year of life and reaches three-fourths of that size by the end of third year. As it enlarges, the brain presses outward on the bony tables and is responsible for the gradual development and general shape of the adult head. When the brain fails to grow properly, the other structures tend to direct their growth inward, thus accounting for the enlargement for the frontal sinus, the increased width of the diploic space and the elevations of the great wing of sphenoid and the petrous ridge on the affected side.[5]

These changes can occur only when the brain damage is sustained before three years of age however; such changes may become evident as soon as nine months.[5]

Conclusion:

A 22 year old male presenting with non convulsive status epilepticus, NCCT and MRI brain revealed hemiatrophy of brain. After ruling out differentials of hemiatrophy of brain, Dyke Davidoff Masson Syndrome was confirmed.

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