



## Case Report

# Dyke-Davidoff Masson Syndrome- A Case Report.

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### ARTICLE INFO

Received 31 Aug. 2014

Received in revised form 09 Sep. 2014

Accepted 16 Sep. 2014

#### Keywords:

Hemiatrophy,

Dyke-Davidoff Masson syndrome.

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### ABSTRACT

A 34 year old male patient was admitted with complaints of the seizure episode in spite of being on anti epileptics. CT scan Brain showed left cerebral hemiatrophy suggestive of Dyke-Davidoff-Masson syndrome (DDMS).

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## Introduction

Dyke-Davidoff-Masson syndrome (DDMS) refers to atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy), which is usually due to an insult to the developing brain during fetal or early childhood period.<sup>1</sup> The clinical features are variable and depend on the extent of brain injury. More commonly they present with recurrent seizures, facial asymmetry, contralateral hemiplegia, mental retardation or learning disability, and speech and language disorders. Sensory loss and psychiatric manifestations like schizophrenia had been reported rarely.<sup>2,3</sup> The typical radiological features are cerebral hemiatrophy with ipsilateral compensatory hypertrophy of the skull and sinuses. The syndrome had been documented mainly in adolescents and adults.<sup>4-6</sup>

## Case Report

A 34 year old male patient was admitted with a history of generalized tonic clonic seizure 2 days back. The patient was a known case of seizure disorder, since last 20 years and was regularly on Tab. Carbamazepine 400 mg BD and Tab. Clobazam 10 mg HS. The patient also had learning difficulties and hence he left his school when he was 10 years old. On examination patient was in the post-ictal confusion. But he had no neurodeficit.

His CT scan Brain was done. It showed left cerebral hemiatrophy, with dilated ipsilateral lateral ventricle, thickened calvarium on the left side and hypertrophied left frontal sinus. These imaging features were suggestive of Dyke- Davidoff- Masson Syndrome (DDMS).

The patient was then started on Tab. Sodium Valproate 300 mg BD and Tab. Clobazam 5mg HS. He did not have any seizure episode during hospitalization and hence discharged. Later he lost follow-up.

## Discussion

In 1933, Dyke, Davidoff, and Masson described the plain skull radiographic and pneumatoencephalographic changes in a series of nine patients characterized clinically by hemiparesis, seizures, facial-asymmetry, and mental retardation.<sup>7</sup> The plain skull radiographic changes included thickening of calvarium and dilatation of ipsilateral frontal and ethmoid sinuses. Also, there was the elevation of the greater wing of sphenoid and petrous ridge and upward tilting of planum-sphenoidale. In 1939, Alpers and Dear defined two types of cerebral hemiatrophy.<sup>8</sup> 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.<sup>9</sup> 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 partly responsible for the gradual enlargement 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 of the frontal sinus, the increased width of the diploic space and the elevations of the greater wing of sphenoid and the petrous ridge on the affected side.<sup>10</sup> These changes can occur only when brain damage is sustained before three years of age however, such changes may become evident as soon as nine-months after brain damage was sustained.<sup>11</sup> It was proposed that a vascular anomaly 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.<sup>12</sup> It was reported that decrease in carotid artery flow due to coarctation of aorta can also cause cerebral hemiatrophy. 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 as in the linear-nevus syndrome. A proper history, thorough clinical examination, and radiologic findings provide the correct diagnosis.

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**Figure 1.** CT scan brain showing left cerebral hemiatrophy, dilatation of left ventricle and thickened left calvarium



**Figure 2.** CT Scan Brain showing hypertrophied left frontal sinus and mastoid air cells