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ABSTRACT:
Dyke-Davidoff-Masson syndrome consists of cerebral hemiatrophy and skull vault changes like hyperostosis and overpneumatization of ipsilateral paranasal sinuses with clinical features of hemiparesis, seizures and mental retardation. We report an adult case of this syndrome in a 37 year old female with crossed cerebellar atrophy as an additional radiologic feature.

Keywords: DDMS (Dyke-Davidoff-Masson syndrome), Hemiatrophy, CT (Computed Tomography)
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INTRODUCTION
Dyke-Davidoff-Masson syndrome is a rare clinical syndrome described by C.G. Dyke, L.M. Davidoff, and C.B. Masson in 1933 [1]. Findings unique to this syndrome are cerebral hemiatrophy, facial asymmetry with compensatory hypertrophy of ipsilateral paranasal sinuses and cranial vault changes caused secondary to brain insult in fetus or childhood [1-3]. It is diagnosed by combination of clinical and imaging features. Although a disease of childhood, the diagnosis of DDMS should be considered as a possibility in adults presenting with long standing history of hemiparesis, seizures and mental retardation [4]. We present a case of this rare syndrome in an adult with associated crossed cerebellar atrophy and discuss about the etiologies, imaging features and differential diagnosis.

CASE REPORT
37-year-old female with left hemiparesis since birth came with history of seizures since last 15 years with recent increased frequency. Patient is on valproic acid for seizures. There is associated speech abnormality and mental retardation since childhood. No prior imaging study was done due to poor socioeconomic status. No family history of seizure disorder present. On Physical examination, left motor weakness with exaggerated tendon reflexes was noted. Blood and biochemistry reports came out normal. No associated skin changes/ port wine stains found on examination.
Plain CT sections revealed hemiatrophy of right cerebral hemisphere with punctate areas of cortical calcification. Dilation of ipsilateral lateral ventricle and subdural collection are noted with falcine displacement to the right. Left cerebellar atrophy with relative preservation of left middle cerebral peduncle is noted. There is overpneumatization of right frontal, right ethmoid and mastoid air cells with thickening of the inner table of cranial vault on right side. Contrast enhanced sections did not reveal any abnormal focal enhancement. Right MCA appears hypoplastic with paucity of distal branches. A diagnosis of DDMS is made based on
clinical history and imaging findings. The patient is kept on regular follow-ups for seizures.

**DISCUSSION**

C.G. Dyke, L.M. Davidoff, and C.B. Masson in 1933 first described this syndrome by demonstrating skull asymmetry, ipsilateral hyperpnuematization and osseous hypertrophy in nine patients with history of hemiparesis, seizures and mental retardation on plain skull x ray and pneumoencephalography. Elevation of sphenoid wings and petrous ridge were addition features [1] Asymmetrical growth of cerebral hemispheres with hemiatrophy was also described as a part of this syndrome. [2, 3]

Clinical features of this syndrome consist of hemiparesis, sensory loss of speech disorder, mental retardation and psychiatric disorders like schizophrenia [3]. A predilection for involvement of male gender and left brain hemisphere has been described [5]. Prognosis is proposed to be better in patients presenting with paresis after 2 years and without recurrent episodes of seizures [3].

The etiology of this syndrome can be due to congenital causes with vascular anomalies in fetus or acquired causes like trauma, infections, vascular and intracranial hemorrhage in perinatal periods [6]. Our case report with above clinical features suggest the congenital type. Multiple vascular anomalies like coarctation in the mid aortic arch between the innominate and common carotid arteries [7] and hypoplastic middle cerebral arteries have been described with the congenital type [8] A study described association of cutaneous vascular anomalies with ipsilateral cortical hemiplasia and multiple intracranial vascular abnormalities like aplasia of internal carotid artery, persistent trigeminal and proatlantal artery and double kinking of internal carotid artery to suggest common developmental anomaly occurring during the same embryonic period [9]. It has been proposed that vascular anomaly in early gestation leads to major brain defects whereas those occurring later leads to localized lesions [10]. Acquired type of this syndrome is less frequently reported with etiologies including encephalitis, [11, 12] febrile seizures, [13] anoxic birth, [14] post cerebral malaria [15] and post stroke. [16] Computed tomography and magnetic resonance imaging are the modalities of choice in assessment of the etiology and extent of cerebral parenchymal involvement [17]
Crossed cerebellar atrophy has been described with this syndrome in few cases in the literature [18-20]. Damage to the corticopontocerebellar pathway is the proposed pathogenic mechanism of crossed cerebellar atrophy [21]. Status epilepticus and extent of supratentorial brain damage were major determinants in development of crossed cerebellar atrophy in patients with long standing destructive brain insults and epilepsy in one study with no role of recurrent seizures [22].

Bony changes associated with this syndrome are seen when brain damage occurs before 3 years of age due to lack of pressure of the growing brain on the bony tables of the skull which leads to inward growth of skull tables and paranasal sinuses [23, 24].

The differential diagnosis for DDMS is Rasmussen encephalitis, Sturge weber syndrome, Fishman syndrome and basal ganglia germinoma [3]. Rasmussen encephalitis is a rare immune mediated brain disorder consisting of cerebral hemiatrophy and intractable seizures. The hemiatrophy in Rasmussen encephalitis is progressive over time [25]. Rasmussen encephalitis differs morphologically from DDMS in absence of skull vault changes [26].

Sturge weber syndrome is a congenital neurocutaneous syndrome with leptomeningeal angiomatosis, nevus flammeus, cerebral lobar atrophy with cortical and subcortical calcifications [27]. Our case report has punctate cortical calcifications but lack the other radiological and clinical features of Sturge Weber Syndrome. Fishman syndrome is a rare congenital neurocutaneous syndrome comprising of unilateral cranial lipomas, lipodermoids of the eye and brain hemiatrophy [28].

The treatment of DDMS is mostly symptomatic with management of seizures, speech abnormalities and learning disabilities. Functional hemispherectomy has been shown to improve prognosis in patients with recurrent seizures [29].

**CONCLUSION**

Dyke-Davidoff-Masson syndrome is a rare syndrome consisting of cerebral hemiatrophy, hypertrophy of ipsilateral paranasal sinuses and cranial vault changes with crossed cerebellar atrophy as an additional feature. Although a disease of childhood, diagnosis should not be missed in adults with typical clinical and imaging features.
CONFLICT OF INTEREST

No conflict of interest exists.

AUTHOR’S CONTRIBUTIONS

The author is involved in diagnosis of the case, follow up and review of literature.

REFERENCES


FIGURE LEGENDS

Figure 1(A) and 1 (B): Plain CT section of the head reveals hemiatrophy of right cerebral hemisphere with punctate areas of cortical calcification. Dilation of ipsilateral lateral ventricle and subdural collection are noted with falcine displacement to the right.

Figure 2: Contrast CT section reveals hypoplasia of Right MCA with paucity of distal branches.

Figure 3 (A) and 3 (B): Bone window reveals overpneumatization of right frontal, right ethmoid and mastoid air cells with thickening of the inner table of cranial vault on right side.

Figure 4: Coronal section reveal crossed cerebellar atrophy involving left side.

FIGURES

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