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Dyke-Davidoff Masson Syndrome: A rare cause of cerebral hemi-atrophy and aggression

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ABSTRACT

Introduction: Dyke-Davidoff Masson Syndrome (DDMS) is a rare neurological disorder characterized by cerebral asymmetry secondary to unilateral brain atrophy, ipsilateral ventricular dilatation, convulsions, mental retardation, learning disabilities and behavioural changes. These patients can potentially have complex medical management that requires multidisciplinary team based approach. Case Report: We describe a case of a 16-year-old female with developmental delay who presented with new onset seizures along with aggressive head-banging behaviour and mild motor weakness in right upper and lower extremeties. Radiological imaging of brain demonstrated left sided cerebral atrophy which was more prominent in both frontal and temporal lobes along with ipsilateral ventricular enlargement, enlarged left sphenoid sinus and prominent left mastoid air cells. Conclusion: Dyke-Davidoff Masson Syndrome (DDMS) is a rare neurological disorder characterized by cerebral hemiatrophy, ventricular dilatation, seizures, behavioural changes and learning disabilities. Patients can have compensatory skull thickening on the affected side along with unilateral frontal and ethmoidal sinus anomalies. Causes of DDMS include cerebral trauma, infections, hemorrhage and ischemia.

Keywords: Cerebral, Hemi-atrophy, Head-banging, Pediatric, Unilateral

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INTRODUCTION

Dyke-Davidoff Masson Syndrome is a rare neurological disorder that can manifest with cerebral hemiatrophy, ventricular dilatation, seizures, hemiparesis, behavioural changes and developmental disabilities [1, 2]. Clinical features can vary depending on the degree of cerebral involvement [1, 2]. It was initially described in 1933 by Dyke D. et al. in a case series of 9 patients who had above mentioned clinical manifestations [3]. Presentation is age dependent since the timing of neurological insult is an important determining factor [1, 2]. Interestingly, pediatric cases are rare since it is most commonly found in adult population [3]. Moreover, it is most commonly found in males with left sided cerebral involvement [4]. Here, we describe a case of a 16-year-old female with a significant past medical history of developmental delay, who presented to us with new onset seizure, aggressive head-banging behavior, mental retardation and characteristic radiological findings of Dyke-Davidoff Masson Syndrome.

CASE REPORT

A 16-year-old female with a significant past medical history of developmental delay presented to us with new onset tonic-clonic seizures and head banging behaviour that began one week prior to admission. As per mother, seizures started with stiffening of upper extremities bilaterally followed by involvement of bilateral lower extremities and foaming around mouth. Last seizure was 7 hours prior to admission which reportedly lasted for two minutes. Head banging behaviour reportedly occurs every 1 to 2 days in addition to progressively worsening aggressive behaviour.

Patient was born via normal vaginal delivery, without any complications at 40 weeks of pregnancy. On physical examination, patient had mild proptosis of left eye along with mild motor weakness in right upper and lower extremities. There was no evidence of microcephaly on gross examination of patient. Blood work including complete blood count, liver function tests and complete metabolite panel was within normal limits. Urine drug screen was negative. MRI (Magnetic Resonance Imaging) of brain revealed left sided Fronto-Temporal cerebral atrophy with widening of sulci and atrophy of gyri (red arrow heads in Figure 1A and Figure 1B); Left ventricular dilatation without midline shift (red asterisks in Figure 1B). Axial CT (Computerized Tomography) scan of brain with contrast revealed asymmetric spheno-ethmoidal recesses, ethmoidal sinuses and prominence of left mastoid air cells (red circles in Figure 2). No evidence of subdural or epidural hematoma was found on imaging. Consequently, diagnosis of Dyke-Davidoff Masson Syndrome was made and treatment was initiated. Initial seizure control was acquired by intravenous Lorazepam 4 mg/kg followed by oral Valproic Acid 400 mg twice per day. Patient was found to be seizure free in outpatient follow ups every two weeks after treatment along with resolution of aggression and improvement in right sided motor weakness. In addition, patient is currently compliant with regular outpatient physical and occupational therapy with improvement in quality of life.

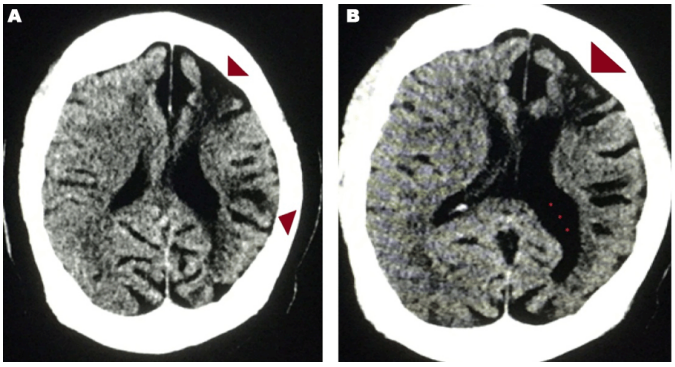

Figure 1: (A) Demonstrates left sided Fronto-Temporal cerebral atrophy with widening of sulci and atrophy of gyri as shown by red arrow heads. (B) MRI demonstrates left sided Fronto-Temporal cerebral atrophy (red arrow heads) along with significant left ventricular dilatation without midline shift as shown by red asterisks.

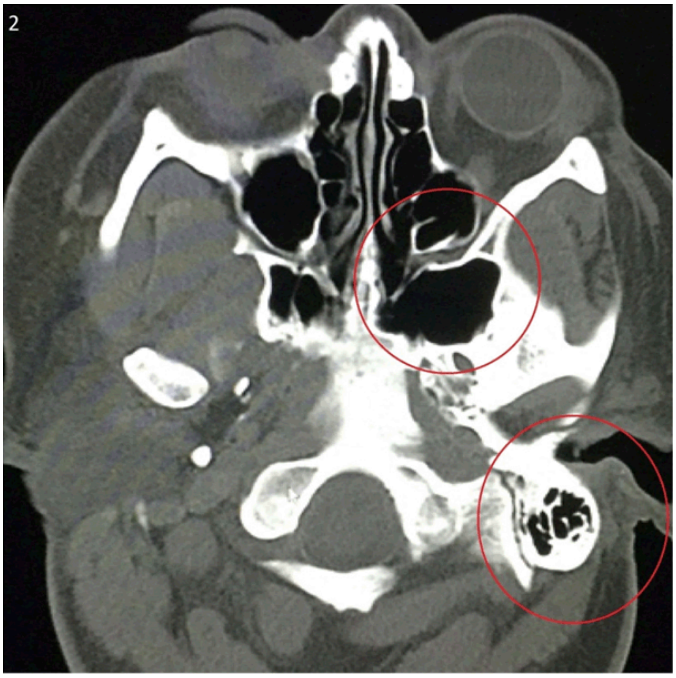

Figure 2: Axial CT scan of brain with contrast revealed asymmetric spheno-ethmoidal recesses, ethmoidal sinuses and prominence of left mastoid air cells (red circles in Figure 2). No evidence of subdural or epidural hematoma noted.

DISCUSSION

Clinical presentation of DDMS varies amongst patients and heavily depends on timing of cerebral insult during neurological development [5, 6]. Earlier insults, during embryological development, tend to present with hemi-atrophy of brain along with compensatory calvarial thickening on the affected side. In addition, patients can have varying degree of learning difficulties, speech disturbances and behavioural problems depending on degree of cerebral involvement [5, 6]. In this case, atrophy of left frontal lobe could potentially explain aggressive head banging behaviour and developmental delay.

Two major types of DDMS are: congenital and acquired [1, 7]. Congenital DDMS is caused by in-utero insult during embryonic development most commonly secondary to intrauterine hypoxia due to gestational vascular occlusion, vascular malformations and infections during first trimester of pregnancy [1, 7]. Due to lack of blood supply to specific cerebral hemispheres during neural development, there is onset of sulcal prominence along with midline shift towards the affected side from the unaffected cerebral hemisphere [1, 7]. On the other hand, acquired causes include birth trauma hypoxia, intracranial haemorrhage, brain tumours, high frequency febrile seizures and vascular causes like aortic arch anomalies [1, 7]. Previous studies have demonstrated MRI and CT scans to be the best imaging modalities to assess neuroanatomical defects in DDMS patients [8]. In a study by Atalar M. et. al., interesting radiological findings like increased prominence of paranasal sinuses, atrophy of cerebral peduncles and thalamus along with

hypoplastic lentiform nucleus and middle cranial fossa were noted [8].

During clinical evaluation, other similar syndromes need to considered in differential diagnosis (Table 1) [7]. These diagnosis were considered and ruled out in this case due to absence of their characteristic findings.

Table 1: Other diseases with similar presentation

Syndrome	Characteristic Features
Basal Ganglia Germinoma	Brain tumor with progressive hemiparesis and hemi-atrophy of brain.
Sturge-Weber Syndrome	Port-wine facial nevus, intracranial calcifications, no midline intracranial shift, seizures, mental retardation and hemiparesis.
Silver-Russell Syndrome	Triangular face, poor bone age, normal intelligence and hemi-hypertrophy of body.
Fishman Syndrome	Intracranial lipoma with ocular changes.
Linear Nevus Syndrome	Facial nevus, recurrent seizures, ventricular dilatation.
Rasmussen Encephalitis	Focal epilepsy with cognitive defects mostly following a viral infection.

Medical management of DDMS is focused on seizure control with suitable anti-convulsive pharmacotherapy [5, 8, 9]. Patients have better prognosis in absence of status epilepticus and seizure onset after two years of age [4, 9]. In addition, multidisciplinary team based approach by enrolling patients in physiotherapy, speech therapy and occupational therapy can improve their functional capacity and quality of life by enabling them to perform ADLs (Activities of Daily Living) independently. In treatment resistant cases, neurosurgical intervention like hemispherectomy are available at select institutions globally [4, 9]. It is known to resolve seizures and hemiparesis in treatment refractory cases [4, 9].

CONCLUSION

Dyke-Davidoff Masson Syndrome (DDMS) is a rare neurological disorder characterized by cerebral hemiatrophy, ventricular dilatation, seizures, behavioural changes and learning disabilities. Patients can have compensatory skull thickening on the affected side along with unilateral frontal and ethmoidal sinus anomalies. Causes of DDMS include cerebral trauma, infections, hemorrhage and ischemia.

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Author Contributions

Smit P. Shah – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Pooja P. Shah – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Cristina Vardi – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor of Submission

The corresponding author is the guarantor of submission.

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Consent Statement

Written informed consent was obtained from the patient for publication of this case report.

Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.