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Case Report

Dyke-Davidoff-Masson syndrome: A rare case of hemiatrophy of brain—Case report from Nepal ☆☆☆

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ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological disorder that results from brain injury during intrauterine or early years of life. Prominent cortical sulci, dilated lateral ventricles, cerebral hemiatrophy, hyperpneumatization of the sinus, and compensatory hypertrophy of the skull are the characteristic findings. We describe a female patient who presented with a history of seizure, right-sided body weakness, and neuroimaging features of left cerebral hemiatrophy, dilatation of left lateral ventricle, left frontal sinus hyperpneumatization, asymmetric calvarial thickening, and elevation of the petrous ridge.

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Introduction

Atrophy or hypoplasia of one cerebral hemisphere (hemiatrophy) is referred to as Dyke-Davidoff-Masson syndrome (DDMS), first described in 1933. This condition is typically brought on by an injury to the developing brain during prenatal or early childhood [1]. The clinical characteristics vary according to the severity of the brain injury. Recurrent seizures, facial asymmetry, contralateral hemiplegia, mental retardation

or learning difficulty, and speech and language difficulties are the most frequent symptoms that they display [2]. Left hemisphere engagement and the male gender are frequently observed. Age of manifestation is dependent on when the neurologic insult occurred, and distinctive alterations may not appear until puberty. The gold standard for the diagnosis of DDMS is computed tomography or magnetic resonance imaging. The underlying pathologic processes' nature and extent vary widely [1]. The radiological finding includes cerebral hemiatrophy, which is associated with ipsilateral lateral ventricle dilatation and prominent sulcal spaces while thalamus

Abbreviations: DDMS, Dyke-Davidoff-Masson Syndrome; MRI, magnetic resonance imaging; CT, computed tomography.

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☆☆ Patient Consent: Consent from patient's father was taken on written form for case report and using MRI images of her daughter in any journal after explaining on his own language.

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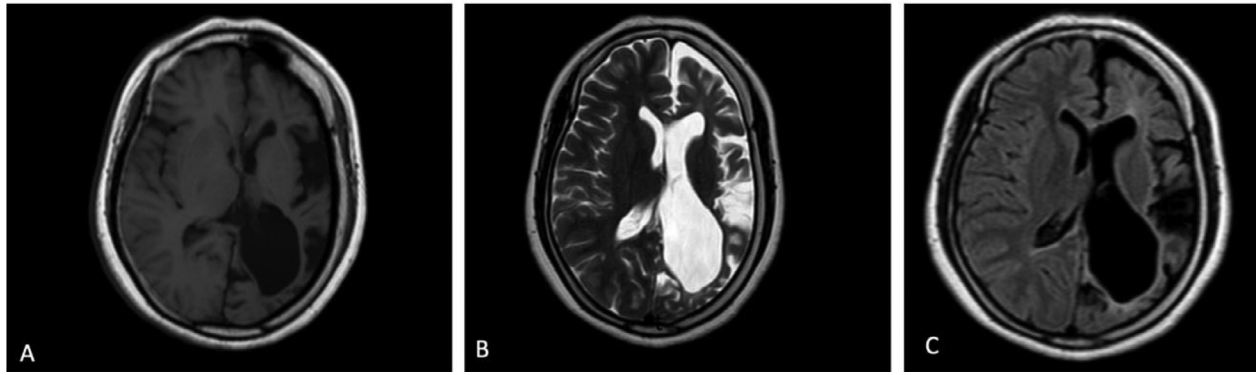


Fig. 1 – (Axial MRI images): (A) T1-weighted, (B) T2-weighted, and (C) FLAIR; demonstrating atrophy of left cerebral hemisphere with dilatation of left lateral ventricle and prominent extra-axial CSF spaces on the left side.

and brainstem atrophy can also be seen but are uncommon [3]. Due to rarity of the disorder, it may be misdiagnosed and under-reported by many clinicians.

Case report

Here we report a case of a 13-year-old female, third child of the family, delivered by vaginal delivery at home. There was no history of hospital visits during antenatal period by her mother. She presented to Neuro-medicine OPD, BIR hospital with a history of weakness in right half of her body which started since her childhood. Her father reported that she has been experiencing episodes of generalized tonic and clonic seizures with brief periods of loss of consciousness since her childhood and increased in frequency over 4 months. She had delayed developmental and intellectual impairment since childhood. No history of surgery, trauma, or significant illness in the past. There was no history of similar presentations in rest of her family members.

On general examination, the patient was irritable with irrelevant talks and abnormal behavior. On neurological workup, power was 3/5 on the right side with decreased sensation and hypertonic right limb. We could also observe a mild facial tilt towards the right. Her blood counts, ESR, liver and renal function tests were within normal limits. The patient had no significant medical history in the past.

Magnetic resonance Imaging (MRI) was done, which revealed the decreased volume of the left cerebral hemisphere with gross dilatation of the left lateral ventricle (hydrocephalus ex-vacuo) demonstrated in (Fig. 1) On GRE sequences there was no evidence of blooming artifact suggesting hemorrhage or calcification. There was calvarial thickening on the left side with hyperpneumatization of the left frontal sinus as compared to the right side, there was also elevation of the petrous ridge on the left side (Fig. 2). Similarly, MRI of the patient revealed atrophy of the midbrain on the left side, probably due to Wallerian degeneration and there was also atrophy of the right (contralateral) cerebellar hemisphere with prominence of folia (Fig. 3). A diagnosis of DDMS was made. Now she

is under anti-epileptic medications but she still experienced seizure episodes.

Discussion

Dyke, Davidoff, and Masson documented 9 patients with pneumato-encephalographic abnormalities on a skull radiograph who had hemiparesis, facial asymmetry, seizures, and mental retardation in 1933 [4].

The case presented here displayed the characteristic radiological and clinical findings that are consistent with a diagnosis of DDMS.

The etiopathogenesis may result from acquired factors such as trauma, infection, vascular abnormalities, and intracranial hemorrhage during the perinatal period or shortly afterward, which cause hemi cerebral atrophy. Vascular insult during intrauterine life may cause hypoplasia of a cerebral hemisphere. These variables, in turn, lead to a decrease in the production of brain-derived neurotrophic factors, which promotes cerebral atrophy [5].

The atrophic hemisphere's large sulci and encephalomalacia are signs of late-onset brain damage brought on by aberrant neuronal and glial growth or apoptosis during cortical development, indicating acquired causes. On the other hand, no noticeable sulci will be present if the brain injury occurs during embryogenesis when the creation of gyri and sulci is incomplete [6].

A precise diagnosis and the implementation of suitable management are made possible by imaging via CT and MRI, which is of great importance. Because they offer cross-sectional images with tiny slices and post-processing capabilities, these 2 imaging modalities are useful. Cerebral hemiatrophy/hypoplasia, hyperpneumatization of the paranasal sinuses, and compensatory osseous hypertrophy are relevant imaging characteristics for DDMS. As the patient ages, these radiological traits will become increasingly noticeable [7].

This disorder has to be distinguished from Rasmussen encephalitis, Basal ganglia germinoma, Sturge-Weber syndrome, Silver-Russel syndrome, Linear nevus syndrome, and Fishman syndrome [2]. Rasmussen encephalitis is a per-

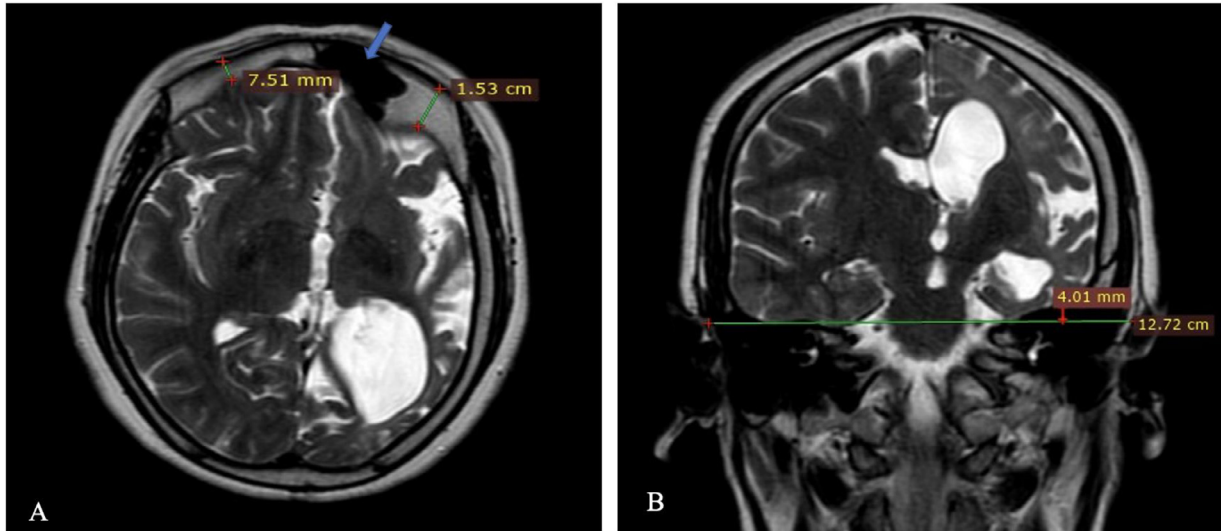


Fig. 2 – (A) Axial T2-weighted image demonstrating calvarial thickening of left frontal region (calvarial thickness measuring 7.15 mm on right side and measuring 15.3 mm on left side and hyperpneumatization of frontal sinus of the left side (arrow on A). **(B)** Coronal T2-weighted image demonstrating elevation of the petrous ridge of the left side.

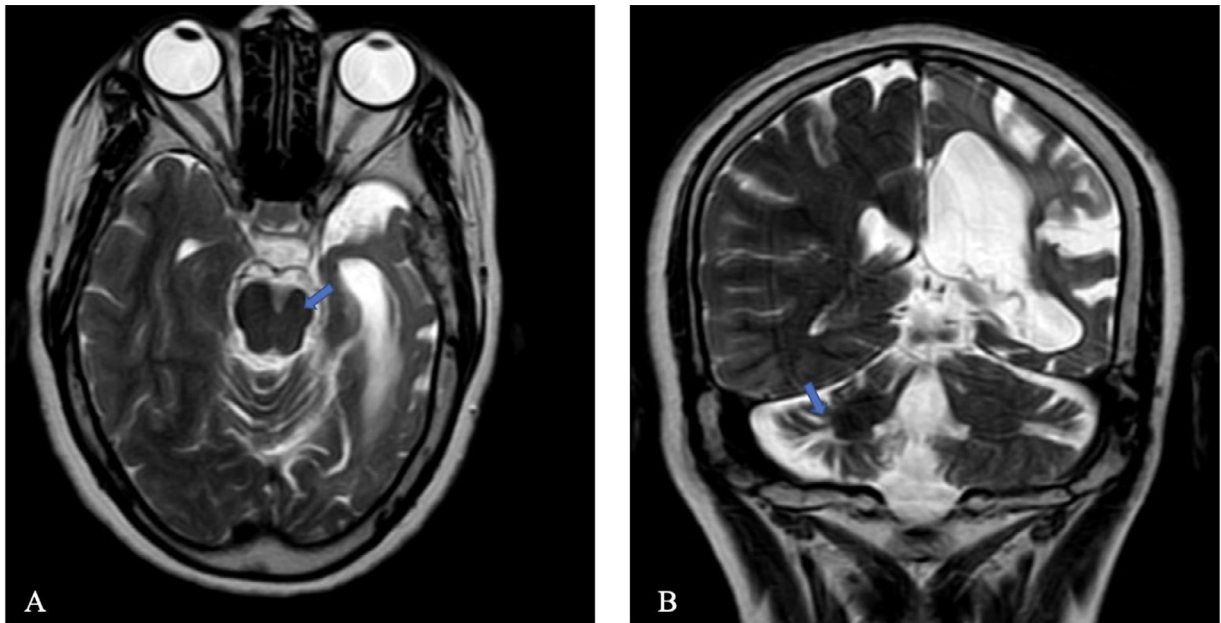


Fig. 3 – (A) T2-weighted axial image demonstrating atrophy of midbrain in left side, due to Wallerian degeneration (arrow). **(B)** Coronal T2-weighted image demonstrating atrophy of right cerebellar hemisphere with prominent cerebellar folia (arrow).

sistent, immune-mediated condition that is hypothesized to develop as a result of viral infections. Intractable focal epilepsy and cognitive deficits in childhood are the typical symptoms. Unilateral hemisphere atrophy without apparent calvarial alterations is one of the imaging characteristics. Leptomeningeal angioma-related brain atrophy is represented by Sturge-Weber syndrome (encephalotrigeminal angiomatosis). The intracranial tram track calcification, the port-wine facial nevus, and the lack of midline displacement

are the identifying characteristics. Whereas, poor growth, delayed bone age, clinodactyly, normal head circumference, normal IQ, the distinctive facial phenotype (triangular face, broad forehead, short pointed chin, and narrow-wide mouth), and hemihypertrophy are all characteristics of Silver-Russel syndrome [2].

Rasmussen encephalitis is unlikely, as the patient had an ipsilateral thickened calvarium with a non-progressive pattern of seizure and neurologic deficit. The patient did not have

skin lesions which are typically seen in Sturge-Weber syndrome which rules out this condition. In the presented case, the patient had no history of trauma, infection, or tumor to suggest an acquired variety of the syndrome.

Since the majority of patients with this disorder present with refractory seizures, management focuses on controlling seizures with the proper anticonvulsants. Additionally, home-based occupational, speech and physical therapy are also important [5]. Patients with hemiplegia and uncontrollable, incapacitating seizures should consider hemispherectomy, which is successful in 85% of instances. Patients with hemiparesis that develops after age 2 or without recurring seizures may have a better prognosis [3]. It is crucial for a rural neurologist or pediatrician to diagnose the condition early using appropriate imaging (CT), as hemispherectomy is not even available in many urban tertiary care centers. Then, the focus of treatment should be on achieving the best seizure control possible while occasionally adjusting medication dosages and performing home physical therapy, especially in a resource-poor country like Nepal.

Conclusion

DDMS is a rare neurological disorder that leads to refractory seizures along with a spectrum of disabilities. Due to the rarity of this condition, it can be easily missed or underreported. A thorough history and imaging can give an early diagnosis and helps to differentiate the condition from other similar conditions. Treatment is supportive with physiotherapy, occupational therapy, speech therapy, and surgery for intractable seizures. Clinicians should be aware of the signs and symptoms, risk factors, and diagnostic features of DDMS

so that the patients could be diagnosed early and managed properly.

Patient consent

The patient's father has given written consent for this case report and for the use of MRI images of his daughter in any article.

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