CASE REPORT

Dyke-Davidoff-Masson Syndrome with Recurrent/Refractory Seizures: A Rare Case in Adult

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Received: 30 November 2022; Accepted: 26 December 2022

ABSTRACT

A 26-year-old left-handed female presented in the emergency ward with three to four episodes of convulsion, which started focally with secondary generalization. She had froth through the mouth, tongue bite, and mild postictal confusion for 1 day before hospitalization. She had a past history of stroke with hemiplegia right 23 years back with partial recovery. About 3 months after hemiplegia, she also had seizures, and since then taking medications for seizures. She had mild mental retardation with poor scholastic performance and left school after the fourth standard. On computer tomography (CT)—brain evaluation, he was found to have imaging features of Dyke-David off-Masson Syndrome (DDMS). It is one of the rare causes of recurrent and refractory seizures in adults.

INTRODUCTION

Dyke-Davidoff-Masson Syndrome (DDMS) presents as seizures, hemiplegia or hemiparesis, and mental retardation. However, mental retardation not always be present. The classical radio imaging feature described is hemiatrophy of the cerebrum, ventricular dilatation, and calvarial changes along with or without shift of midline structure to the affected side depending on whether brain insult was in utero or after the birth of a baby. This was first described by Dyke–Davidoff–Masson in 1933.

Since then, only a few cases of DDMS have been reported in late childhood and adults.2–4

CASE DESCRIPTION

A 26-year-old, left-handed female presented in the emergency ward with three to four episodes of recurrent convulsion starting on the right upper and lower limb, followed by secondary generalization. She had froth through her mouth, tongue bite, and mild postictal confusion since 1 day before hospitalization at our institute. She had no history of headache, neck pain, vomiting, loss of consciousness, and fever. No clear history of any prenatal or postnatal insult. Past history of hemiplegia on the right side 23 years back, which recovered partially. About 3 months after hemiplegia, she had a history of seizures that were recurrent, and she has been taking multiple anti-convulsants since then. She also had a history of mild mental subnormality with poor scholastic performance and left her study after the fourth standard. She was receiving tab valproate 200 mg twice a day (BD), tab carbamazepine 200 mg BD, tab phenytoin 100 mg three times daily (TDS), and tab frisium 10 mg HS along with folinic acid 5 mg once daily. There was no CT-brain report available of her childhood hemiplegia. Clinically her hemodynamic and vital parameters were stable. Both carotids were felt and equal. No sebaceous nodules over the face, no nevus, no ash-leaf rash, or shagreen patch over a body part. She had right supranuclear facial palsy and hemiparesis with 4/5 power in the upper limb and 3/5 in the lower limb. Deep tendon reflexes on the right side were exaggerated, and the plantar was extensor. The left side was normal. The coordination and sensory examination were normal. Cardiovascular, respiratory, and abdominal system examination was normal. A routine investigation like complete blood count, kidney function test, liver function tests, serum sodium, serum potassium, and serum magnesium was normal. Serum calcium was 6.1 mg/dL (reduced). TDM level of valproate was reduced to 26.47 (50–100). CT-brain revealed left cerebral atrophy and ventricular dilatation with no shift of midline structures which is suggestive of DDMS (Fig. 1). CT-angiography reveals no vascular occlusion or malformation. Along with the correction of calcium, we gave her intravenous (IV) levipril 500 mg TDS, IV valproate 500 mg TDS, oral carbamazepine through RT 200 mg TDS, and tab. Frisium 10 mg Hs along with folic acid 5 mg daily. With this treatment also, for the first 3–4 days, she used to get two to three episodes of seizures, and then on the 5th day, she became seizure-free. Subsequently, her injectable drugs were started orally. Now with four oral antiepileptic, calcium, and folic acid, she is doing well on follow-up.

Fig. 1: Computed tomography (CT) brain showing left cerebral atrophy with right lateral ventricular dilatation with no shift of midline

Discussion

The first description of DDMS dates back to 1933 when Dyke–Davidoff–Masson described the plain skull radiographic and pneumoencephalographic change in a series of nine patients presenting with hemiparesis, seizures, facial asymmetry, and mental retardation.1 DDMS is caused by the cerebral insult that may occur in utero when the maturation of calvarium has not been completed or during early life (during birth or postnatally first 2–3 years) due to brain damage (usually traumatic).5 The etiological factors for DDMS have been postulated as trauma, inflammation, vascular malformations, or occlusion.2–4

When the insult occurs in utero, there is a shift of midline structures towards the side of the disease, and the sulcal prominence replacing the gliotic tissues is absent.6 This feature differentiates it from cerebral hemiatrophy, which occurs in early life. The atrophied cerebral hemisphere will have

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prominent sulcal spaces if the insult occurs after birth or after the end of sulcation.\footnote{7} But in our patient, we were not able to postulate the exact cause for hemiatrophy as CT-angiography was normal, and there is no clear history of prenatal or postnatal insult.

When the insult occurs \textit{in utero}, it could be due to gestational vascular occlusion primarily involving the middle cerebral artery (MCA) territory. Decreased carotid artery blood flow due to coarctation of the aorta can also cause cerebral hemiatrophy.\footnote{3} Garg et al.,\footnote{6} reported febrile seizures as a possible etiological factor for cerebral hemiatrophy, while MCA stroke by Sener and Jinkis.\footnote{7}

Approximately one-third of patients with seizures/epilepsy present as refractory, requiring multiple antiepileptic drugs.\footnote{8} Juvenile myoclonic epilepsy, Lennox–Gastaut syndrome, tuberous sclerosis, and bilateral cerebral palsy are some of the disorders which can present with mental retardation and refractory seizures. DDMS is one of the rare conditions presenting with recurrent/refractory seizures. Proper clinical history and CT-magnetic resonance imaging (CT-MRI) will provide a correct diagnosis. Inadequate therapy and precipitating factors like hyponatremia, hypocalcemia, hypomagnesemia, and hypoglycemia should also be considered in refractory seizures, as in our patient, she had hypocalcemia and a low therapeutic level of valproate. We corrected hypocalcemia and increased the dose of valproate, and subsequently, she responded.

\textbf{Dyke-Davidoff-Masson Syndrome (DDMS)} should be differentiated from basal cell germinoma, Sturge–Weber syndrome, linear nevus syndrome, Fishman syndrome, Silver–Russel syndrome, and Rasmussen encephalitis.\footnote{7} Proper clinical history and CT-MRI findings will help in clinching the correct diagnosis.

The treatment is symptomatic and should target convulsion, hemiplegia/hemiparesis, and learning difficulties. The prognosis is better if hemiparesis occurs after the age of 2 years and in the absence of prolonged or recurrent seizures. Intractable disabling hemiplegia or seizures are the potential candidates for hemispherectomy, with a success rate of 85\% in carefully selected cases.\footnote{10} 

\textbf{Dyke-Davidoff-Masson Syndrome (DDMS)} should be suspected in adults who have a past history of hemiparesis due to vascular, traumatic, or infective causes during early childhood. Radio imaging with CT- or MRI-brain usually reveals characteristic features of DDMS.

\textbf{References}