



Unusual Presentation of Dyke-Davidoff-Masson Syndrome

Authors

Dr Deepak Agrawal¹, Dr Manish Khandelwal², Dr Nilesh Kumar Patira³

¹ADMO (Physician), Central Hospital, North Western Railway, Jaipur, India

²Assistant Professor, Dept of Anaesthesia & Critical Care, RUHS College of Medical Sciences, Jaipur, India

³Assistant Professor, Dept of Medicine, American International Institute of Medical Sciences, Udaipur

Abstract

Background: *Dyke-Davidoff-Masson syndrome (DDMS) is a rare disease which is clinically characterized by hemiparesis, seizures, facial asymmetry, and mental retardation. The classical radiological findings are cerebral hemiatrophy, calvarial thickening, and hyperpneumatization of the frontal sinuses. This disease is a rare entity, and it mainly presents in childhood. Adult presentation of DDMS is unusual and has been rarely reported in the medical literature. We report a Case of DDMS in a 37yr old female who presented with Tonic Clonic Seizures involving right half of the body with secondary generalization and Spastic Hemiparesis of right half of the body with facial asymmetry without mental retardation.*

Material and Methods - Nil

Result - Nil

Conclusion: *This case report shows an unusual presentation of the rare Dyke-Davidoff-Masson syndrome (DDMS) in an adult female.*

Keywords: *Dyke-Davidoff-Masson syndrome, spastic hemiparesis, seizures.*

Introduction

First description of Dyke-Davidoff-Masson syndrome (DDMS) dates back to 1933, when Dyke, Davidoff and Masson described the plain skull radiographic and pneumatoencephalographic changes in a series of nine patients. Since then only few pediatric cases of DDMS have been reported. The classical findings including cerebral hemiatrophy along with calvarial thickening and hyperpneumatization of the frontal sinuses are only found if an insult to the brain occurs before 3 years of age. The major concern of the disease remains the intractable seizures for which drug therapy is not sufficient in most of the cases, and a

surgical approach is necessary. However, if the patient presents later in life, the presentation may not be similar to that seen in childhood, and management changes accordingly.

Case Report

A 37-year-old woman presented to our outpatient department with recurrent Tonic Clonic Seizures involving right half of the body with secondary generalization and Spastic Hemiparesis of right half of the body with facial asymmetry. Birth history was indicative of a full-term normal delivery without any antenatal or perinatal complications. She had normal developmental

milestones during infancy till 6 years of life. At the age of 6 years she developed febrile seizures for which she was treated but since then she continued to have seizures which exacerbated due to poor compliance. She got married at the age of 20 years and conceived after 5 years of married life and delivered a healthy male child without any complications but since then seizure frequency increased and she was advised not to have pregnancy in future. She also has slurred speech, facial deviation during episodes. Her seizures did not respond to several antiepileptic medications in different combinations. Her examination did not reveal any neurocutaneous markers and had a right-sided spastic hemiparesis with brisk tendon reflexes and extensor plantar response. A magnetic resonance imaging (MRI) of the brain was subsequently done which revealed left cerebral hemiatrophy; ipsilateral dilatation of lateral ventricles, ipsilateral midline shift, hypoplasia of thalamus, with ipsilateral pneumosinus dilatans (frontal). Ipsilateral left fronto-parietal hyperostosis was also seen. We accordingly kept a diagnosis of DDMS and managed her conservatively with muscle relaxants like baclofen and physiotherapy, carbamazepine 200 mg three times a day and clobazam 10 mg at bed time as well as counseling, after which her symptoms improved.

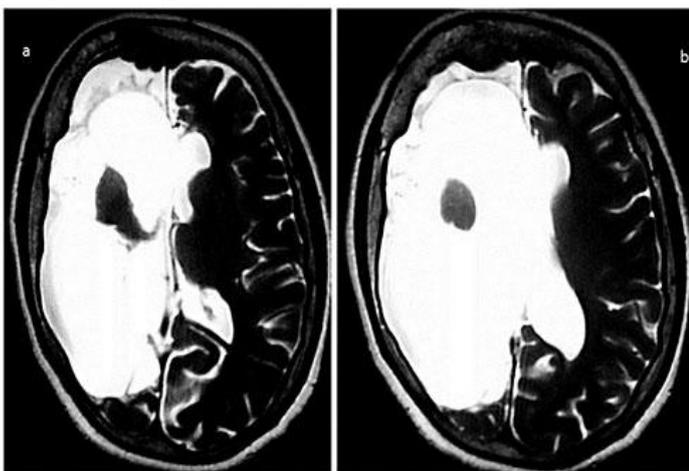


Fig.1 - T2-weighted axial MRI showing cerebral hemiatrophy on the right side of the brain at the level of the basal ganglia (a) and at the supraganglionic level (b).

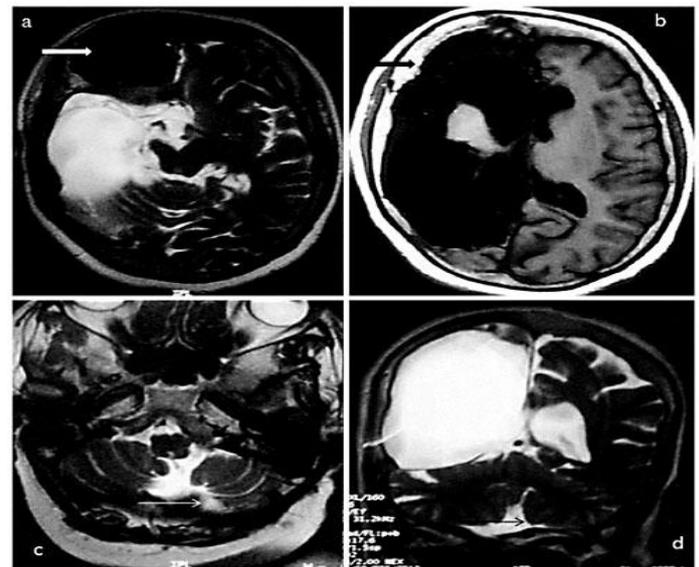


Fig.2 - MRI of the brain showing frontal sinus hypertrophy in an axial T2-weighted sequence (thick white arrow; a) along with calvarial thickening in an axial T1-weighted sequence (thick black arrow; b). Crossed cerebellar hemiatrophy is evident on the left side in axial T2-weighted (thin white arrow; c) and coronal T2-weighted images (thin black arrow; d).

Discussion

This rare condition derives its name from the researchers Dyke, Davidoff, and Masson who first reported the condition way back in 1933. They described plain skull radiographic changes in 9 patients who presented with seizures, facial asymmetry, hemiparesis, and mental retardation. Subtotal or diffuse cerebral hemiatrophy is a classical imaging finding. However, unilateral focal atrophy may occasionally be noted in the cerebral peduncles and the thalamic, pontine, crossed cerebellar, and parahippocampal regions. Brain imaging may additionally reveal prominent cortical sulci, dilated lateral ventricles and cisternal space, calvarial thickening, ipsilateral osseous hypertrophy with hyperpneumatization of the sinuses (mainly frontal and mastoid air cells), and an elevated temporal bone. The clinical features include contralateral hemiparesis with an upper motor neuron type of facial palsy, focal or generalized seizures, and mental retardation along with learning disabilities. There is no sex predilection, and any side of the brain can be

involved, although involvement of the left side and male gender has been shown to be more common in one study.

However mental retardation was not always present and seizures may appear months or years after the onset of hemiparesis. The etiological factor for Dyke-Davidoff-Masson syndrome has been postulated as trauma, inflammation or vascular malformations and occlusions. When the insult occurs in-utero, it could be due to gestational vascular occlusion, primarily involving the middle cerebral vascular territory. A possible etiological relation of cerebral hemiatrophy and seizures has been reported by different studies in India. Dyke Davidoff Masson Syndrome should be differentiated from Basal cell germinoma, Sturge Weber syndrome, Linear Nevus syndrome, Fishman syndrome, Silver Russell syndrome and Rasmussen encephalitis. A proper clinical history and CT/MRI findings provide the correct diagnosis. The treatment is symptomatic, and should target convulsion, hemiparesis and learning difficulties. Prognosis is better if hemiparesis occurs after the age of 2 years and in absence of prolonged or recurrent seizures. Children with intractable disabling and hemiplegia are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases.

This patient had the acquired variety of the disease as the patient's complaints started after an episode of febrile seizure. Although initially the patient had progressive symptoms in the form of intractable seizures, later, in spite of a progressive loss of dexterity, the frequency of seizures decreased, which is atypical for this condition, as the classical presentation of this disease itself is intractable seizures. However, the natural course of this disease in adults has not been described in detail earlier due to the paucity of adult presentation; nevertheless, there have been pediatric case reports where, despite progression of the disease in the form of hemiatrophy of the brain and hemiparesis, the seizure frequency was dramatically reduced.

As previous imaging was not available in our patient, it is difficult to know with certainty whether the hemiatrophy originated from childhood or whether there was any progression of atrophy later in life; however, the decrease in the seizure frequency points towards a progressive atrophy later.

Conclusion

For DDMS cases presenting in early childhood, refractory seizures remain the usual concern. Accordingly, hemispherectomy is the treatment of choice with a success rate of 85% in selected cases. However, if the presentation is late as in our case and if seizures are under control, the patient can be kept on antiepileptic medications in spite of surgery, along with supportive therapy including physiotherapy, speech therapy, and occupational therapy. Further longitudinal studies are required to ascertain the natural course of this syndrome especially in an adult population, which would help in planning strategies regarding the time and nature of interventions and management accordingly.

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Conflicts of interest: There are no conflicts of interest

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