Acquired Dyke-Davidoff-Masson syndrome (DDMS)

Jitender Aneja, Satyarth Jangli¹, Manpreet Singh¹, Amit Mittal²

Department of Psychiatry, Government Medical College and Hospital, Chandigarh, Haryana and Punjab, ¹Departments of Psychiatry and ²Radiology, Maharishi Markandeshwar Institute of Medical Sciences and Research, Ambala, Haryana, India

INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) is an uncommon congenital or acquired neurological syndrome named after the doctors who reported a series of nine patients having hemiplegia associated with cranial asymmetry on skull x-rays in 1933.[1] It is characterized by cerebral hemiatrophy, contralateral hemiplegia, calvarial thickening, skull and facial asymmetries, cognitive impairment, and epilepsy. The diagnosis of this uncommon and interesting syndrome has been made in 6,000-year-old skeletal remains (from the skull),[2] in utero,[3] infancy,[4] childhood,[5,6] and late adulthood.[7-9] Patients with DDMS commonly present with neurological manifestations like seizures that may be partial or generalized, aphasia, sensorineural hearing loss (SNHL), and hemiparesis. So, the diagnosis of this syndrome is largely based on the clinical and radiological findings. The etiology of the congenital (infantile) variant is unclear; however, intrauterine vascular occlusion has been hypothesized to be the possible cause.[8] The causal factors of acquired DDMS may be trauma, infection, vascular abnormality, ischemia or hemorrhages, coarctation of the aorta, subependymal germinial matrix, amniotic bands, and intraventricular hemorrhage of premature infants.[1,4,6,10] Most of the available literature is in the form of case reports[1-16] and a few case series,[17,18] where the clinicoradiological features and neurocognitive manifestations have been described in detail. Some of the patients were reported to have refractory seizures[8,16] but the associated neuropsychiatric symptoms like hemiparesis, hearing loss, aphasia, mental retardation, and cognitive deficits further contribute to a poor prognosis.

A comprehensive literature review showed many cases of congenital and early onset of DDMS. However, acquired variants of DDMS with an infectious etiology are rarely reported. Adding to the literature, we report a case of acquired DDMS with classical neuroimaging findings and a range of neuropsychiatric symptoms.

CASE REPORT

A 10-year-old boy presented with recurrent right-sided focal seizures with secondary generalization and behavioral abnormalities. He was born of nonconsanguineous marriage. There were no ante/perinatal complications and he was delivered vaginally at full-term at a primary health-care center. There was no history of delayed milestones and no behavioral abnormalities till the age of 6 years when he developed
high-grade fever followed by seizures. Seizures started with the right upper limb focal tonic-clonic posturing followed by secondary generalization associated with frothing from the mouth, tongue bite, urine, and fecal incontinence. Subsequent to seizure, he did not regain consciousness for the next half an hour and was taken to the local hospital. He was treated there for 2 days but had a recurrence of seizures with regaining of consciousness in between the episodes. So, he was referred to a tertiary care center in Northern India where he was hospitalized with a working diagnosis of viral meningoencephalitis made on the basis of the presentation with high-grade fever and seizures, despite his cerebrospinal fluid (CSF) examination being within normal limits. The latter may be explained by the inadvertent delay in the analysis of CSF samples, which could lead to cell destruction in CSF and thus, a normal examination. However, serological examination and neuroimaging were not done during hospital stay. As per the available medical records, he was treated with intravenous ceftriaxone and acyclovir, with which fever and seizures remitted. He was discharged with neurological sequelae in the form of profound bilateral SNHL (clinically and on pure tone audiometry) and aphasia. He also had severe behavioral problems in the form of unprovoked anger outbursts, aggression, socially disinhibited behavior, loss of previously learnt adaptive skills, hyperactivity, and impulsivity. He needed continuous supervision of a family member for his hygiene and safety. The family did not seek any medical treatment, thereafter, till he had seizures again at the age of 9 years 9 months. There was no change in the semiology of seizures. He had four such seizures since the time of relapse with aggravation of behavioral problems, and was unmanageable at home. Hence, he was brought to the outpatient psychiatry clinic of this rural tertiary care hospital located in Northern India.

He was uncooperative during his physical and mental state examinations. He did not speak on his own, was very fidgety, and did not sit at one place. There were no signs of extrapyramidal syndrome or any abnormal movement disorders. He was unable to comprehend the commands given for the examinations that could be due to a combination of hearing impairment and agitation, but we did not utilize nonverbal methods to assess his comprehension. His neurological examination revealed brisk reflexes in the upper limbs. The patient did not cooperate for performance-based intelligence tests; so, his intelligence quotient (IQ) was calculated based on the Vineland Social Maturity Scale (VSMS). His mental age was found to be that of a 5-year-old with a social quotient of 50. A diagnosis of organic brain syndrome (postencephalitic) and epilepsy were considered and he was further investigated.

The blood biochemistries and hemogram were within normal limits. The electroencephalograph (EEG) showed asymmetrical background slowing in the left-sided leads with intermittent spike and sharp transients arising predominantly from the left temporal leads. The magnetic resonance imaging (MRI) of the brain showed left cerebral hemiatrophy with prominent sulcal spaces with area of gliosis [Figure 1- dashed arrow] and dilated left ventricle [Figure 1- solid arrow]. The T2-weighted (T2W) fluid-attenuated inversion recovery (FLAIR) image [Figure 2] showed cerebral atrophy with prominent temporal horn (solid white arrow) and calvarial thickening (dashed white arrow). There was hyperintensity seen in the right cerebellar hemisphere [Figure 3] on the T2W image.

Although, crossed cerebellar atrophy (CCA) is not a commonly reported finding in DDMS syndrome, it has been described previously. The underlying pathogenesis may involve damage to the corticopontocerebellar tract or centrolobular cerebellar sclerosis due to prolonged seizure effect.[19,20] It had also been suggested that the extent of supratentorial lesion and the antecedent of status epilepticus, but not recurrent seizures, might be related to the development of CCA.[21,22] So, on the basis of clinical and radiological findings, a diagnosis of DDMS was entertained. The patient was started on carbamazepine with an initial dosage of 200 mg/day in divided dosages that were increased to 800 mg/day in divided dosages due to poor control of seizures, along with augmentation with clobazam of 10 mg/day and olanzapine of 5 mg/day (mouth dissolving tablets) for aggressive behavior. The seizures were controlled and behavioral symptoms reduced by nearly 50%, as reported subjectively by the family. We further plan to assess the patient for nonpharmacological management of behavioral problems.

The magnetic resonance imaging (MRI) of the brain showed left cerebral hemiatrophy with prominent sulcal spaces with area of gliosis (dashed arrow) and dilated left ventricle (solid arrow)

![Image](image-url)
DISCUSSION

Dyke, Davidoff, and Masson first described this syndrome based on the radiological findings. The present literature search up to October 2014 shows more than 60 reported cases of DDMS with varying features. However, acquired cases of DDMS of possible infectious etiology have been reported infrequently. In the index case, an acquired etiology is considered on the basis of uneventful child birth, normal development of milestones, and no evidence of neuropsychiatric manifestations till he developed fever and seizures. The radiological findings of prominent sulci and lesser bony changes also suggest an acquired pathology. So, the etiology of DDMS in the present case may be meningoencephalitis and febrile seizures, which possibly inhibited development of the affected cortex. The behavioral manifestations might be due to disruption of the orbitofrontal circuits (due to atrophy of the left frontal lobe), thus leading to aggression and irritability. The differential diagnoses considered on the basis of shared clinicoradiological features of seizures, hemiparesis, mental retardation, facial asymmetry, cerebral hemiatrophy, and calvarial changes were Rasmussen’s encephalitis, Sturge-Weber syndrome, Silver-Russell syndrome, linear nevus sebaceous syndrome, progressive multifocal leukoencephalopathy (PML), Fishman syndrome, and basal ganglia germinoma. Sturge-Weber syndrome, linear nevus sebaceous syndrome, and Fishman syndrome are neurocutaneous syndromes with characteristic facial cutaneous malformations and intracranial vascular malformations, which were not present in the index case. Silver-Russell syndrome is a chromosomal disorder with early onset (congenital), characteristic facial and skeletal dysmorphisms, and growth retardation. Intracranial tumors like germinoma and PML have peculiar radiological features other than cerebral hemiatrophy, which differentiate them from DDMS. Rasmussen’s encephalitis is a chronic, progressive, immune-mediated brain disease, which has close resemblance to clinical and radiological features of DDMS. The onset is abrupt with polymorphism of seizures with frequent occurrence, intractability, and persistent motor seizure activity, i.e., epilipsia partialis continua. It was the closest differential in the index case but was ruled out on the basis of radiological findings. However, rare but unilateral cerebral hemiatrophy due to viral encephalitis has been known to occur. The classical radiological findings and clinical manifestations help in the diagnosis of DDMS. The treatment of this uncommon disorder is largely symptomatic in the form of adequate control of seizures and behavioral manifestations. Functional hemispherectomy has been shown to be good in those with refractory seizures. It has also been seen that prognosis may be better in patients in whom hemiparesis appeared after 2 years of age or in whom the seizures were not recurrent. Although it is not necessary to do neuroimaging in every pediatric case presenting with febrile seizures, it should be done in those who also have neurobehavioral manifestations, changes in adaptive skills, and frequent breakthrough seizures so that one does not miss out on the diagnosis.

CONCLUSION

In conclusion, the present case illustrates the classical clinical and neuroimaging findings of DDMS and brings the attention of clinicians to this uncommon syndrome.

ACKNOWLEDGEMENT

We thank Dr. Ankush Sharma, MD, DM (Neurology), Assistant Professor, Department of Medicine, Maharishi Markandeshwar Institute of Medical Sciences and Research (MMIMSR) for his valuable inputs.
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How to cite this article: Aneja J, Jangli S, Singh M, Mittal A. Acquired Dyke-Davidoff-Masson syndrome (DDMS). Int J Adv Med Health Res 2015;2:55-8.
Source of Support: Nil, Conflict of Interest: None declared.