Dyke Davidoff Masson Syndrome Presenting with Intellectual Disability with Behavioral Problems and Substance Use Disorder: A Case Report

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SUMMARY

Dyke Davidoff Masson Syndrome (DDMS) is a rare condition with varied presentation. Characteristic features are cerebral hemiatrophy, enlargement of ipsilateral ventricle, enlargement of ipsilateral air sinuses and clinical finding of contralateral hemiparesis or hemiplegia and seizures. DDMS may have comorbid intellectual disability or speech disorder but presentation with psychiatric disorders is rare. We present a case of DDMS who first came to attention with behavioral problems associated with aggression, social disinhibition and with alcohol and cannabis use disorder.

Keywords: Atrophy, paresis, hemiplegia, aggression, substance use disorder

INTRODUCTION

The etiology of Dyke Davidoff Masson Syndrome (DDMS) is not uniform. It may be because of antenatal cause, difficult delivery or infection or trauma after birth. The clinical picture may also vary and is usually characterized by facial asymmetry, seizure, cerebral hemiatrophy, contralateral hemiparesis or hemiplegia, intellectual disability etc. Investigations may show dilatation of homolateral ventricle, thickening of skull and expansion of air sinuses on the affected side (Dyke et al.1933).

Though several cases of DDMS have been reported from different parts of the world, presentation with psychiatric manifestations is rare. We are reporting a case from the North Eastern part of India, which is the first from this region. The patient presented with intellectual disability, behavioral problems and alcohol and cannabis use disorder.

CASE REPORT

A 38-year-old unmarried male was brought to the psychiatry out-patient department by his family members, who accounted for the six-year history of the patient with socially disinhibited behaviour, defiant attitude, excessive anger and aggressive behaviour, such as destroying household articles or physically attacking others, including children in the family and the neighborhood. These episodes of anger and physical aggression were mostly precipitated when his demand for money was not met by his elder brother which he spent on alcohol and cannabis. His family had not observed symptoms suggestive of alcohol withdrawal when the patient did not have access to any of the substances for days. The patient's brother reported that the patient demonstrated socially inappropriate behavior at times, such as watching pornography even in the presence of others or attempting intimacy with a domestic help despite being cautioned after her complaints. These behaviors were seen even when the patient was not under

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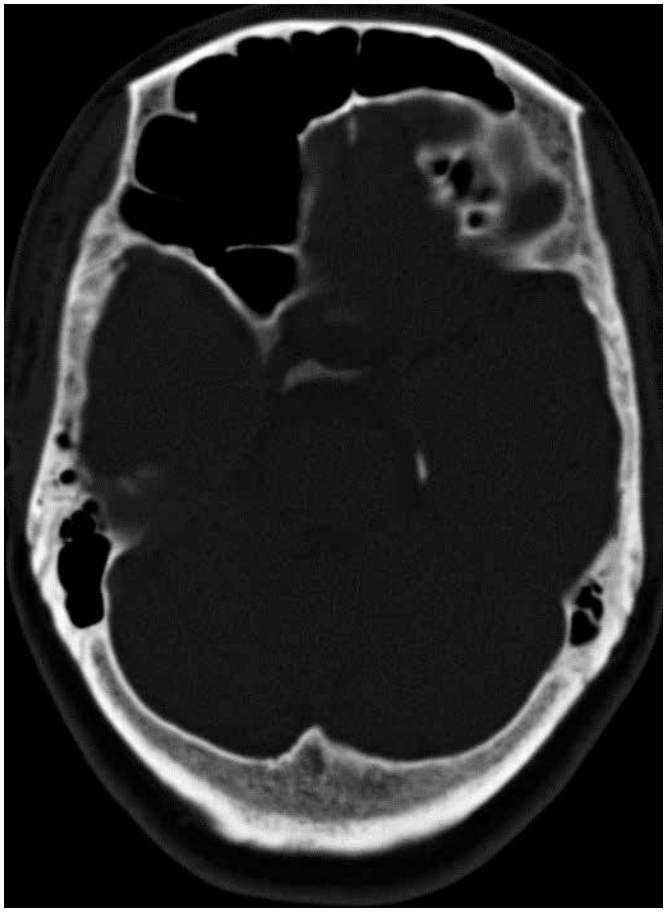

Figure 1: Axial CT images showing hypertrophied frontal sinus and mastoid air cells on right side.

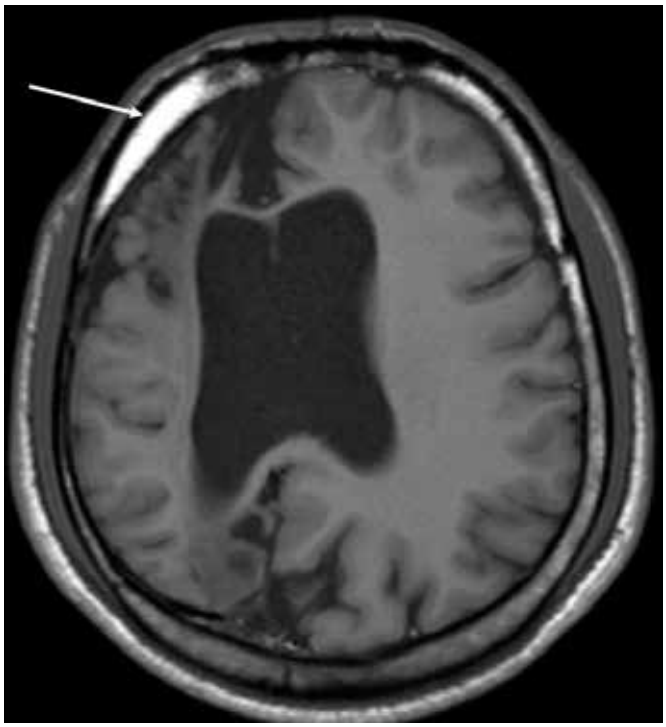

Figure 2: T1W axial image showing atrophy of right cerebral hemisphere, calvarial hypertrophy on right side (arrow)

Figure 3: T2W coronal image showing gliosis , atrophy of right cerebral hemisphere, dilated right lateral ventricle. Also to note elevated right petrous ridge (solid arrow)

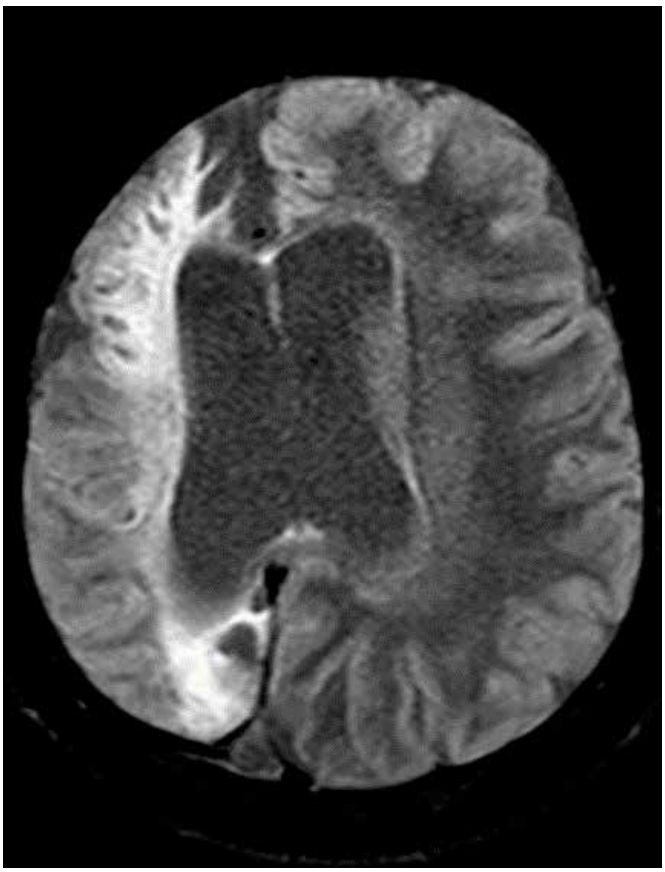

Figure 4: Axial FLAIR image showing gliosis of right cerebral hemisphere.

the effect of alcohol or cannabis. The family did not report any symptoms suggestive of eating or sleep disorder. Early developmental history included delayed milestones. Patient had a history of generalized tonic-clonic seizure (GTCS) which started at the age of 6 years and remitted spontaneously by 8 years of age. Since then, there was no episode of GTCS. He also had a history of non-progressive weakness of the left side of the body since the age of 8 years. His academic performance had been poor, ending by dropping out of school after failing repeatedly in the sixth grade. Before this hospital admission, he was helping his brother in his grocery shop in mostly manual works.

During his examination the patient was conscious and oriented. He was found to have upper motor neuron type of hemiparesis, hypertonia and hyper-reflexia all on the left side and left sided plantar response. He walked with slight difficulty, but his gait was steady without ataxia. His Intelligence Quotient (IQ) was found to be 51 which correspond to Mild Mental Retardation. Magnetic Resonance Imaging (MRI) revealed gliosis and atrophy of the right cerebral hemisphere with dilatation of the right lateral ventricle. There was hypertrophy of the frontal sinus and thickening of frontal, mastoid and occipital bone on the right side (Fig 1-4). The diagnosis of DDMS with Mild Intellectual Disability with Alcohol and Cannabis use Disorder was made. Electroencephalogram (EEG) recorded before admission in the ward did not show any abnormality. After admission, he and was started on Lorazepam 2mg at bed time and tablet Acamprosate 1998 mg in three divided doses. No signs of substance withdrawal were noted by the psychiatrist during the hospital stay. His hemogram revealed normal counts. His liver function and kidney function tests were within normal limits. Drug toxicology screening for different drugs was not done. During the hospital stay, he was not very communicative during the mental status evaluation. He was cooperative with the hospital staff and would comply with the instructions given for self-care and medication. However, because of his hostile behavior towards his family members, Sodium Valproate 1gm was added just before discharge. Post discharge the compliance was poor. Later only Risperidone 2mg was started and was continued on which he showed good control of aggression. Episode of seizure or socially inappropriate behavior was not reported till the last follow up.

DISCUSSION

Dyke et al.(1939) reported a series of nine cases presenting with clinical picture of infantile hemiplegia with X-ray images showing thickening of the cranial vault on the side of cerebral atrophy, over development of frontal and ethmoid air sinuses and of the pertrous part of the temporal bone on the affected side. Other findings were dilatation of the lateral ventricles on the side of the lesion. The etiological causes of cerebral hemiatrophy can be primary or congenital and secondary or acquired. In primary cases, the cerebral injury is mainly vascular in origin and the symptoms appear at or soon after birth. In secondary cases, the cerebral insults occur during the perinatal period or afterwards (Sener and Jinkins 1992). Shen et al. (1993) described three patterns of cerebral hemiatrophy based on MRI findings: Pattern I, diffuse cortical and sub cortical atrophy; Pattern II diffuse cortical atrophy associated with expanded porencephalic cysts; Pattern III: old infarction with necrosis in the territory of the middle cerebral artery. In the case discussed here, the well-formed sulcal spaces points that the vascular insult occurred after birth or after the completion of sulcation which suggest Pattern III.

The differential diagnosis of DDMS includes Sturge-Weber Syndrome, Silver Russel Syndrome, Rasmussen's Encephalitis which can be ruled out clinically along with radiological evidences.

In Sturge-Weber syndrome, the intracranial pathology includes vascular anomaly in the form of leptomeningeal angiomatosis. Clinically facial cutaneous vascular malformation (port-wine nevus) is found affecting the upper face usually in the distribution of the ophthalmic division of the trigeminal nerve. Other associated clinical findings include seizures, transient stroke like neurological deficits, headache and glaucoma. Hemiparesis, hemiatrophy may occur contralateral to the cortical abnormality. Computed Tomography usually shows calcifications in meningeal arteries and veins underlying the leptomeningeal anastomosis. It is common to find cerebral atrophy ipsilateral to the leptomeningeal anastomosis (Thomas-Sohl et al. 2004). Silver-Russel syndrome is characterized by low birth weight, prenatal and postnatal growth retardation, short stature, and broad forehead with triangular face, small and narrow chin, clinodactyly of the fifth finger, asymmetrical body and normal intelligence (Qui and Shi 2007). Rasmussen's encephalitis is a rare, progressive and inflammatory disease of the brain affecting one hemisphere and is associated with intractable focal epilepsy, hemiparesis and cognitive decline. The EEG, in the initial stage of the disease shows slowing and multiple epileptogenic anomalies but in the later part it may decrease and shift to the contralateral hemisphere which is actually due to propagation phenomenon (Sheybani et al. 2011). The MRI shows cortical atrophy without calvarial thickening. In our case the EEG was normal and the MRI showed calvarial thickening.

Only few cases of DDMS with psychiatric comorbidity like schizophrenia (Puri et al. 1994) and schizoaffective disorder (Amann et al. 2004) have been reported. However, the case

under discussion is unique in being the first where comorbid substance use disorder is observed. The aggression that the patient usually showed was not acting out behavior which resulted from delusion or hallucination but can be attributed to firstly low frustration tolerance as it was evident only when his demands were not met and secondly to alcohol and cannabis intoxication. De winter et al. (2011) in their systematic review on challenging behavior among people with intellectual disabilities concluded that medical conditions like urinary incontinence was significantly correlated with physical aggression and visual impairment with self-injurious behavior. They did not find challenging behavior among intellectually disabled with epilepsy except in case which had more frequent seizures, with additional visual and motor impairments and/or with generalized EEG activity. None of these was observed in our patient.

Intellectually disabled people are likely to develop substance use disorder because of poor refusal skills (McGillicuddy and Blane 1999). They also tend to binge when they engage in substance use and intoxication amplifies the danger of high-risk behavior by further impairing their already compromised judgement skills. For example, they may get charged legally for indecent behavior or sexual abuse (Cocco and Harper 2002).

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patients understand that his names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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REFERENCES

Amann B, Garcia de la Iglesia C, McKenna P et al (2009) Treatment-refractory schizoaffective disorder in a patient with dyke-davidoff-masson syndrome. CNS Spectr 14:36-9.

Cocco KM, Harper DC (2002) Substance Use in People with Mental Retardation:A Missing Link in Understanding Community Outcomes? Rehabilitation Counseling Bulletin 46:33-40.

De Winter CF, Jansen AA, Evenhuis HM (2011) Physical conditions and challenging behaviour in people with intellectual disability: a systematic review. Journal of Intellectual Disability Research 55: 675-98.

Dyke CG, Davidoff LM, Masson CB (1933) Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. Surgery, Gynecology & Obstetrics 57:588-600.

McGillicuddy NB, Blane HT (1999) Substance use in individuals with mental retardation. Addict Behav 24:869-78.

Puri BK, Hall AD, Lewis SW (1994) Cerebral hemiatrophy and schizophrenia. Br J Psychiatry 165:403-5.

Sener RN, Jinkins JR (1992) MR of craniocerebral hemiatrophy. Clin Imaging 16:93-7.

Shen WC, Chen CC, Lee SK et al (1993) Magnetic resonance imaging of cerebral hemiatrophy. J Formos Med Assoc 92:995-1000.

Sheybani L, Schaller K, Seeck M (2011) Rasmussen's encephalitis: an update. Schweiz Arch Neurol Psychiatr 162:225-31

Thomas-Sohl KA, Vaslow DF, Maria BL (2004) Sturge-Weber syndrome: a review. Pediatr Neurol 30:303-10.

Qui BP, Shi CH (2007) Silver-Russell syndrome: a case report. World J Pediatr 3:68-70.