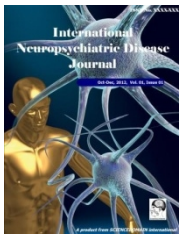

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Dyke-Davidoff-Masson Syndrome- A Rare Cause of Refractory Epilepsy

Prerna Malik[1], Anil kumar D. Gulia[2*], Rajinder Garg[3] and Preeti Singh[1]

[1]Department of Psychiatry, Pt. BDS University of Health Sciences, Rohtak, Haryana, India.
[2]Department of Orthopaedics, BPS Government Medical College, Khanpur Kalan, Sonepat, Haryana, India.
[3]Gian Sagar Medical College and Hospital, Banur, Punjab, India.

Authors' contributions

This work was carried out in collaboration between all authors. Author PM designed the study, wrote the protocol, and wrote the first draft of the manuscript. Author AKDG manuscript preparation, editing and review. Author RG and PS managed the literature searches. All authors read and approved the final manuscript.

Case Study

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ABSTRACT

DDMS is a rare syndrome characterized by seizures, facial asymmetry, contralateral hemiplegia and mental retardation. The characteristic radiologic features are cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. Dyke-Davidoff-Masson Syndrome (DDMS) is one among the syndromes associated with refractory epilepsy. We report a case of DDMS in an 18 year old female who presented with seizures, hemiparesis of the right side and mental retardation. Computed tomography on this patient assisted in making a diagnosis of DDMS.

Key words: Dyke Davidoff Masson syndrome; refractory epilepsy; cerebral hemiatrophy.

1. INTRODUCTION

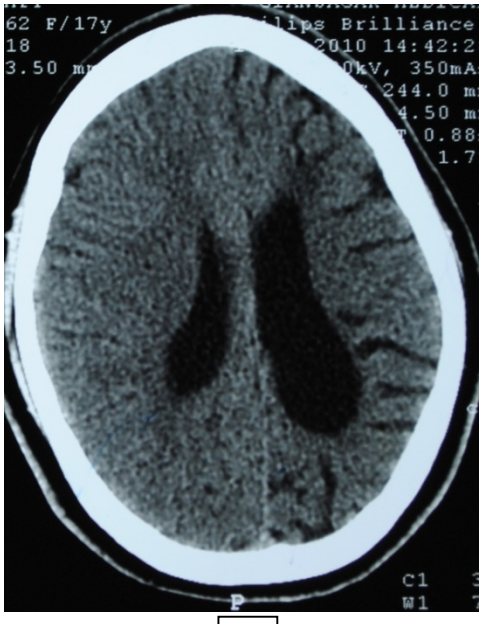
Dyke-Davidoff-Masson Syndrome (DDMS) is one among the syndromes which are associated with refractory epilepsy and was first described by Dyke, Davidoff and Masson in 1933 [1]. They described the plain skull radiographic and pneumatoencephalographic

**Corresponding author: Email: guliaanil@yahoo.com;*

changes in a series of nine patients characterized clinically by hemiparesis, seizures, facial asymmetry and mental retardation. The radiographic features of the skull were asymmetry, ipsilateral osseous hypertrophy of the calvarium and hyper-pneumatization of the sinuses [1]. Since then, there were few case reports in the literature. We are hereby describing the clinical and radiological features of DDMS in a patient with refractory epilepsy.

2. CASE REPORT

An 18 year old girl presented with increasing number of generalized tonic clonic seizures over a six week period. There was history of epilepsy since nine months of age with delayed milestones and she had weakness in right limbs since birth. There was no history of significant antenatal or perinatal complication. There was no family history of epilepsy. She was tried on oral carbamazepine, phenobarbitone and phenytoin at escalating doses since childhood but despite regular and multiple medications, she continued to have occasional breakthrough seizures. She had no new precipitating epileptogenic factors in the recent past. On examination, she was found to be conscious, cooperative and had no nevus on her face nor any signs of meningial irritation. She was submitted for psychometric investigation with Binet-Kamat test of mental ability and her IQ was 62, with mild level of intellectual disability. She had right sided spastic hemiparesis, exaggerated reflexes and an extensor plantar reflex on the right side. Her routine hematological, cerebrospinal fluid examination and biochemical investigations were within normal limits. A plain computed tomography (CT) of head was done which revealed atrophy of the left cerebral hemisphere with dilatation of the ipsilateral lateral ventricle along with low density areas in white matter of left cerebral hemisphere. There is also prominence of ipsilateral sulci as well as mild enlargement of overlying left parietal bone Fig. 1, 2 and 3. So a diagnosis of DDMS was made.



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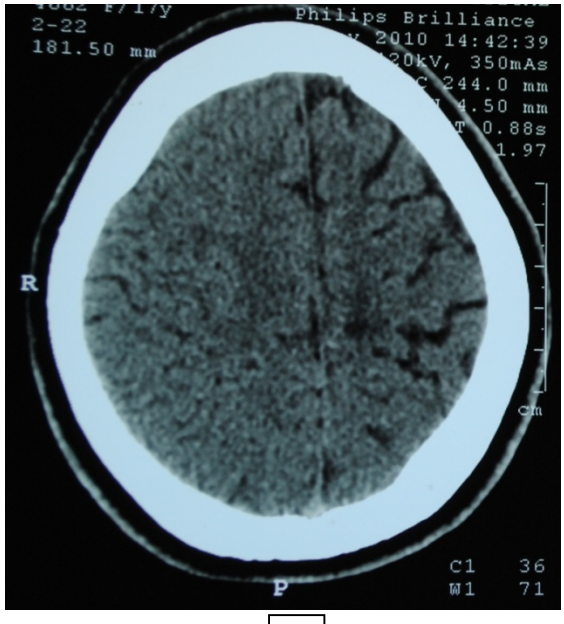

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Fig. 1 & 2. Plain CT images of the brain show left cerebral hemispheric atrophy with left lateral ventricular dilatation and diffuse white matter hypodensities. The falx is seen in the midline with mild thickening of overlying left parietal bone.

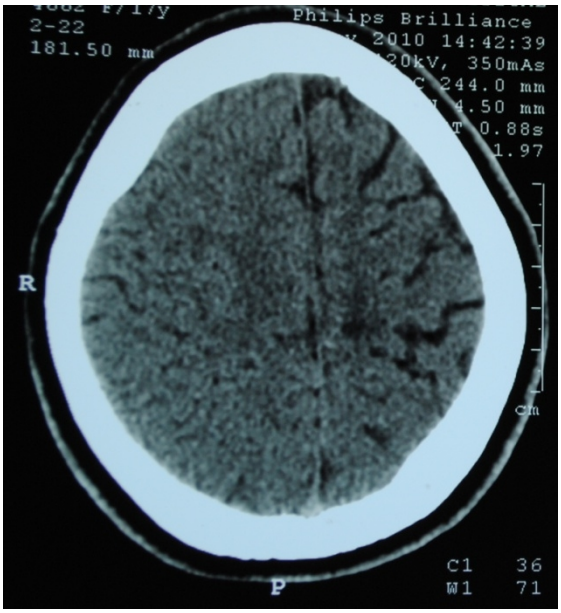

Fig. 3. Plain CT Head showing widening of the sulci and atrophy of gyri of left cerebral hemisphere

The patient is currently receiving valproic acid (1750mg/day) and topiramate (300mg/day). The frequency of seizures has been markedly reduced. Physiotherapy was also started for the weakness of the right sided limbs.

3. DISSCUSSION

The control of refractory epilepsy is a challenge. As such identification of the cause of epilepsy is essential for planning management. There are various tests which have been described to investigate epilepsy. Neuroim aging is one of the main tools used in the investigation of epilepsy. There are many syndromes which are associated with refractory epilepsies and some of them are identified by their imaging characteristics [1].

DDMS is a rare condition characterized clinically by variable degrees of facial asymmetry, seizures, contralateral hemiparesis and mental retardation in association with classical radiological findings of asymmetry of cerebral hemispheric growth with atrophy on one side, ipsilateral osseous hypertrophy and hyperpneumatization of sinuses [1-3]. Cerebral hemiatrophy leads to homolateral hypertrophy of the skull and sinuses. The consequences are facial asymmetry and elevation of the sphenoid wing and petrous ridge [1]. The brain reaches half of its adult size during the first year of life and reaches three-fourths of that size by the end of third year. As it enlarges, the brainpresses outward on the bony tables and is partly responsible for the gradual enlargement and general shape of the adult head. When the brain fails to grow properly, the other structures tend to direct their growth inward, thus accounting for the enlargement of the frontal sinus, increased width of the diploic space and the elevations of the greater wing of sphenoid and the petrous ridge on the affected side. So, these changes can occur only when brain damage is sustained before three years of age [4]. Both sexes and any of the hemispheres may be affected but male gender and left hemisphere involvement are more frequent. Age of presentation depends on time of

neurologic insult and characteristic changes may be seen only in adolescence. The clinical findings may be of variable degree depending on the extent of the brain injury. Varying degrees of atrophy of one half of body, sensory loss, speech and language disorder, mental retardation or learning disability and psychiatric manifestations like schizophrenia may also be present [5]. A proper history, thorough clinical examination and radiologic findings provide the correct diagnosis [6].

The etiology of DDMS may be roughly divided into two categories either congenital or acquired. In the congenital type, the cerebral insult is believed to be vascular in origin and occurs in-utero. In this, the entire cerebral hemisphere is characteristically hypoplastic, there is shift of midline structures towards the side of the disease and the sulcul prominence replacing the gliotic tissue is absent. This feature differentiates it from cerebral hemiatrophy of acquired type. Trauma, infection, vascular abnormalities or intracranial hemorrhage in the perinatal period or shortly thereafter may be responsible for acquired type [7] .The atrophied cerebral hemisphere will have prominent sulcal spaces if the vascular insult occurs after birth or after end of sulcation. Our case seems to be of acquired type. The manifestations of DDMS may be so subtle as to be overlooked on plain radiographs, however, CT/magnetic resonance imaging (MRI) is the diagnostic modality of choice [3]. A possible etiological relation between cerebral atrophy and seizures has been reported in three different studies from India [6,8,9].

The condition needs to be differentiated from basal cell germinoma, Sturge Weber syndrome, Linear Nevus syndrome, Fishman syndrome, Silver-Russell syndrome and Rasmussen encephalitis. Assessment of the complete clinical history and examination along with radiological features can only provide the diagnosis of the Dyke-Davidoff-Masson Syndrome [7,10].

The treatment is symptomatic and should target convulsions, hemiplegia, hemiparesis and learning difficulties. The treatment of DDMS with multiple anti-epileptics is the best option. Children with intractable disabling seizures and hemiplegia are the potential candidates for hemispherectomy with a success rate of 85% in carefully selected cases. Prognosis is better if hemiparesis occurs after the age of 2yrs and in absence of prolonged or recurrent seizures [6].

4. CONCLUSION

In conclusion, DDMS should be considered in a case of refractory seizures with hemiparesis or mental retardation.

CONSENT

All authors declare that written informed consent was obtained from the patient's father for publication of this case report and accompanying images.

ETHICAL APPROVAL

Not Applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Dyke CG, Davidoff LM, Masson LB. Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinus. Surg Gynecol Obstet. 1933;57:588-600.
2. Tasdemir HA, Incesu L, Yazicioglu AK, Belet U, Gungor L. Dyke Davidoff Masson syndrome. Clin Imaging 2002;26:13-17.
3. Sharma S, Goyal D, Negi A, Sood RG, Jhobta A, Surya M. Dyke-Davidoff Masson syndrome. Indian J Radiol Imaging 2006;16:165-166.
4. Parker CE, Harris N, Mavalwala J, Dyke-Davidoff-Masson Syndrome: Five case studies and deductions from dermatoglyphics. Clinical Pediatrics. 1972;11(5):288-292.
5. White JH, Rust JB. Davidoff-Dyke-Masson syndrome presenting as childhood schizophrenia. J Autism Dev Disord. 1979;9:37-40.
6. Narain NP, Kumar R, Narain B. Dyke-Davidoff Masson Syndrome. Indian Pediatr. 2008;45:927-928.
7. Sener RN, Jinkins JR. MR of craniocerebral atrophy. Clin Imaging. 1992;16;93-97.
8. Nair KP, Jayakumar PN, Taly AB, Arunodaya GR, Swamy HS, Shanmugam V. CT in simple partial seizures in children: a clinical and computed tomography study. Acta Neurol Scand. 1997;95:197-200.
9. Garg RK, Karak B. Cerebral hemiatrophy: a possible etiological relation with febrile seizures. Indian Pediatr. 1998;35:79-81.
10. Rao KCVG. Degenerative diseases and hydrocephalus. In: Lee SH, Rao KCVG, Zimmerman RA, editors. Cranial MRI and CT. 4th edition. New York: McGraw-Hill. 1999;212-214.

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