

Case Report

DYKE-DAVIDOFF-MASSON SYNDROME IN A MIDDLE AGED WOMAN

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ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare clinical condition characterized by seizures, mental retardation, facial asymmetry, contralateral hemiplegia. The characteristic radiologic features are cerebral hemiatrophy with compensatory homolateral hypertrophy of the skull and sinuses. We report a case of DDMS in a 38 yrs old female patient who presented with seizures and radiologically atrophy of left cerebral hemisphere with hyper pneumatized ipsilateral frontal sinus.

Key Words: Seizures, DDMS, Adult Patient, Cerebral Hemiatrophy

INTRODUCTION

DDMS was first described by Dyke *et al.*, Davidoff and Masson in 1933 in a series of nine patients with plain skull radiographic and pneumato encephalographic changes (Dyke *et al.*, 1933). Clinically the condition is characterized by seizures, facial asymmetry, contralateral hemiplegia, and mental retardation, behavioural changes like schizophrenia, difficulty and impairment of speech development. Radiologically it is characterised by cerebral volume loss, enlargement of ventricle, prominent sulci, enlargement of paranasal sinuses and skull wall thickening (Hageman *et al.*, 1985, Jacoby *et al.*, 1977). The above features may be present in varying combinations and degrees of severity. The condition is usually diagnosed in childhood but in rare instances, this condition has been diagnosed in teenagers and young adults as well. Here we describe a 38 year old female patient who presented to us with seizures without striking features of neurological deficits but with characteristic radiologic findings suggestive of DDMS.

CASES

A 38 year old Indian female presented to our emergency department with a history of self-fall with a brief period of loss of consciousness following sudden onset of giddiness. This episode was associated with up rolling of eyeballs, tongue bite and no evidence of faecal or urinary incontinence as reported by the bystander. On regaining consciousness, she was oriented with no residual weakness of limbs. She had experienced a similar episode the day before presentation to the hospital. On further probing, she gave a history of an episode of seizure during her pregnancy 10 years ago, for which she received treatment without any radiological investigations. But shortly later on, she had discontinued the medications on herself.

Birth history of the patient revealed that she was born of a full term normal vaginal home delivery with no complications during or after labour as reported by her mother. No history suggestive of head trauma, central nervous system infection during her infancy or childhood was obtained. Her mother was not able to recollect the appropriate time when her daughter went through different developmental milestones. There was no family history of seizure disorder. She was diagnosed to have Hanson's disease (confirmed by nerve biopsy) seven months ago and was on regular treatment.

On systemic examination, minimal facial asymmetry to right side, prominence of naso labial fold, expressive dysphasia, recent memory loss and cortical sensory impairment were detected. She also had difficulty in concentration and attention, disabilities in learning to read (dyslexia), difficulty in calculation (dyscalculia) and difficulty in writing (dysgraphia). She preferred to use her left hand in her day-to-day

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activities except for food consumption and writing. The right side limb was 1 cm thinner than the left side at all levels of examination.

Investigations showed normal electrolytes, blood sugar, urea, creatinine and liver function test. Complete blood count showed normocytic normochromic anaemia with a mildly elevated ESR of 48 mm. Computerized Tomography of the brain with contrast showed gliosis with encephalomalacia involving left frontal lobe and insula with unilateral atrophy in frontal and parietal lobes and prominence of sulci with hyperpneumatized ipsilateral frontal sinus (Figure 1, 2, 3).

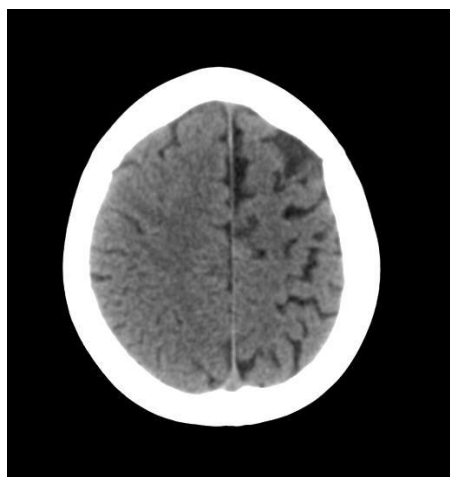


Figure1: Computerized Tomography of the brain showing unilateral dilatation (left side) of cortical sulci with ipsilateral parenchymal volume loss.

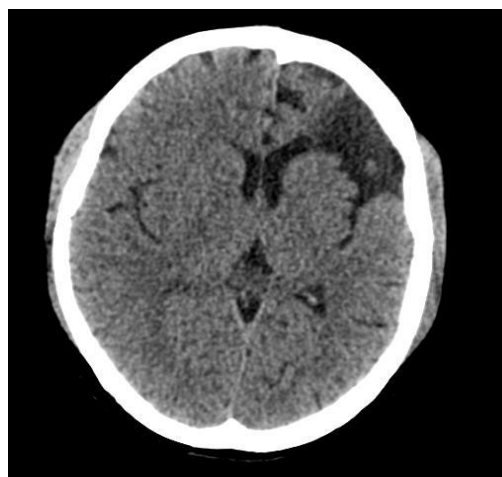


Figure 2: Computerized Tomography of the brain showing dilatation of sylvian fissure with ipsilateral dilatation of ventricle.

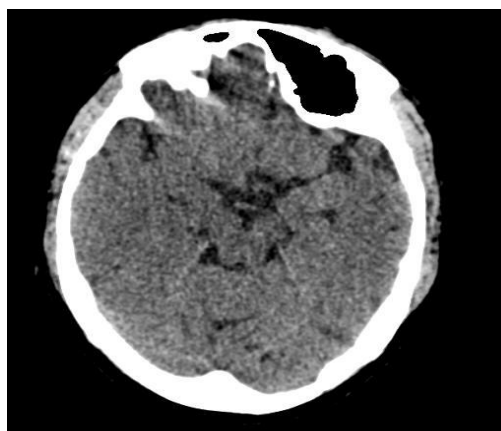


Figure 3: Computerized Tomography of the brain showing enlargement of left frontal sinus

A provisional diagnosis of DDMS was made and the patient was started on regular antiepileptic treatment. The patient had no further episodes of seizure during the hospital stay and was discharged. The patient is presently doing well on regular follow up.

DISCUSSION

DDMS is characterised by variable degrees of unilateral loss of cerebral volume, contralateral hemiplegia, and compensatory changes of calvarium. The etiology of DDMS may be divided into two categories,

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either congenital or acquired. In the congenital type, the various causes propounded are gestational or neonatal vascular occlusion involving the middle cerebral vascular territory, unilateral cerebral arterial circulation anomalies, coarctation of the mid aortic arch, mesencephalon hypoplasia and Wallerian degeneration. Possible pathogenesis in congenital type is lack of cerebral development which results in cerebral hemi-hypoplasia rather than atrophy (Nurisener and Randy, 1922).

Congenital type of DDMS usually presents in perinatal or infancy period with seizure, mental retardation and/or weakness. The age of presentation depends on time of insult and the characteristic changes may be seen only in adolescence (Hageman *et al.*, 1985). There are only few cases of congenital type of DDMS in the adult age group reported in literature to date. In our patient, the developmental history was reported to be normal by her mother. Interestingly, she was asymptomatic for a period of twenty six years, after which she had developed the first episode of seizure during her pregnancy. Since she had poor understanding about her clinical problem, she had discontinued her treatment and was not evaluated further. Ten years later, now she has presented with seizures and after evaluation found to have congenital type of DDMS. To the best of our knowledge our patient is the oldest patient diagnosed with DDMS without gross motor deficits.

In congenital type of DDMS the radiological features are hemiatrophy associated with compensatory ipsilateral osseous hypertrophy and hyper pneumatization of paranasal sinuses. When the cerebral development is poor in-utero or during first two years of life, it is associated with certain cranial changes like ipsilateral hypertrophy of the skull and sinuses as a compensatory change to take up the relative vacuum created by the hypoplastic cerebrum. Radiologically our patient had classical features described in congenital type of DDMS like gliosis with encephalomalacia involving left frontal lobe with unilateral atrophy in frontal and parietal lobes and hyper pneumatization of left frontal sinus.

Cerebral hemiatrophy with seizures could be due to the Sturge-Weber Syndrome, brain tumours, Rasmussen's encephalitis or Silver-Russell Syndrome. However, assessment of the complete clinical history and examination along with radiological features could only provide the diagnosis of the Dyke-Davidoff-Masson Syndrome (Pendse *et al.*, 2004; Shetty *et al.*, 2003).

Acquired type of DDMS results from trauma, infection, ischemia and haemorrhage. As the patient gave no history of previous hospital admissions for prolonged illness or trauma and no evidence of ischemia or haemorrhage found on radiological imaging, the possibility of acquired DDMS was ruled out.

The treatment for DDMS is symptomatic and includes management of seizures, hemiplegia or hemiparesis. The prognosis for the disease is better for patients who develop hemiparesis beyond the age of two years and also if there is no history of prolonged or recurrent seizures. Accordingly, our patient was started on phenytoin and educated on the nature and prognosis of the disease. The prognosis in our patient is expected to be good as she developed seizures only in the second decade and that she was able to carry out her daily activities and functioning.

Conclusion

In conclusion, our case is one among the rare presentations of congenital type of DDMS with seizures and functional defects of frontal and parietal cortices like dyslexia, dysgraphia and dyscalculia. The delayed clinical presentation and the elusive clinical features in our patient were probably due to milder form of the disease. However the radiological features were classical of congenital type of DDMS. This highlights the need for adequate radiological investigations in addition to detailed history and thorough clinical examination in such patients to arrive at the final diagnosis.

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