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Case Report Open Access

Dyke-Davidoff-Masson Syndrome: A Cause of Recurrent Seizures

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Abstract

Objective: Dyke-Davidoff-Masson Syndrome (DDMS) is one of the rare causes of recurrent seizures and can be missed in adults. It is associated with contralateral hemiplegia, mental retardation and facial asymmetry. Confirmed diagnosis can be made with characteristic radiological picture. Hemi-spherectomy is the treatment of choice.

Case: DDMS is usually diagnosed in childhood. Here a rare case of refractory epilepsy is being reported in a 23 years old pregnant woman which was diagnosed as DDMS on radiological findings of cerebral atrophy, ventricle enlargement, dilatation of sulci and osseous hypertrophy of calvarium with increased width of diploic space.

Conclusion: A proper history, thorough clinical examination and characteristic radiological findings provide the correct diagnosis of DDMS. CT/MRI must be performed in patients presenting with recurrent seizures.

Keywords: DDMS; Cerebral hemiatrophy; Recurrent seizures; Refractory epilepsy

Introduction

The most common cause of recurrent seizures is attributed to epilepsy. Though refractory epilepsy is common in neurology practice, Dyke-Davidoff Masson syndrome (DDMS) is one of the rare syndromes associated with it. DDMS includes mainly neurologic symptoms, such as seizures, facial asymmetry, contralateral hemiplegia, and mental retardation. Radiologic study reveals cerebral hemiatrophy, ventricle enlargement, shift to the affected side, dilation of sulci, and compensatory skull changes [1].

DDMS is usually diagnosed in childhood but rare cases have been reported in teenagers and adults [2-4]. We are reporting an interesting case of refractory epilepsy in a 23 years old pregnant woman who was later diagnosed as a case of Dyke-Davidoff Masson syndrome (DDMS).

Case Report

A 23-years-old primigravida, married for two years, presented to our department with seven months pregnancy and recurrent episodes of seizures. On examination, the patient was mentally subnormal but conscious to her surroundings. Her blood pressure was normal and urinary proteins were negative. She had facial asymmetry and right sided hypertonic exaggerated reflexes. Per Abdomen examination revealed 28 weeks size relaxed uterus with cephalic presentation and regular fetal heart rate. There was a history of fits earlier for which she had taken irregular treatment. A provisional diagnosis of seizure disorder (epilepsy) was made. Patient was put on injectable phenytoin which is used as first line of drug for and she responded. After 2 days, she again had seizures and injectable sodium valproate was added. After a seizure free period of one week, patient was discharged on oral medication of phenytoin and sodium valproate. She reported again with seizures after 3 days and on detailed evaluation, it was found that she was not compliant. Injectable sodium valproate and phenytoin were restarted and the patient responded. Her routine haematological and biochemical investigations were within normal limits. Sonographic assessment of foetus was normal. After one week, she developed mild pre-eclampsia with blood pressure of 150/96 mm/Hg and 1+ urinary protein. Due to prematurity, conservative management was continued along with 6 hourly BP charting and foetal monitoring.

At 32 weeks of gestation, the patient complained of loss of fetal movements. Her blood pressure was found to be 160/110 mm Hg with 2+ urinary proteins. Per abdomen examination revealed 34 week size tender and tense uterus with absent foetal heart sounds. Ultrasound examination confirmed intra-uterine death. Diagnosis of severe pre-eclampsia with abruption was made. Bishop's score was 5/13. Artificial rupture of membranes obtained haemorrhagic liquor. Labour was augmented with oxytocin. Patient did not deliver even after ten hours of augmentation and cervical dilatation did not improve beyond 5cm. Caesarean section was performed in view of abruptio placentae with non-progress of labour and a dead female child weighing 2 Kg was delivered. Intra-operative period was uneventful. Post-operatively, patient had seizures which were not controlled with injectable phenytoin and sodium valproate. She became disoriented and irritable. On enquiring repeatedly, patient's mother revealed the fact that she was a known case of seizure disorder since 10 years of age when she had sudden onset of generalised tonic clonic seizures and right sided hemiplegia. She recovered with medication but right sided weakness and facial asymmetry persisted. Since childhood, patient was taking phenytoin irregularly.

CT scan of head showed left cerebral atrophy, ventricle enlargement, dilatation of sulci and osseous hypertrophy of calvarium with increased width of diploic space on left side (Figure 1). A diagnosis of Dyke-Davidoff-Masson syndrome was made. Neurologist opinion was sought who started injection phenobarbitone. Seizures were controlled but the Glasgow coma scale remained 6/15 may be due to the added complication of pregnancy induced hypertension. The patient developed disseminated intravascular coagulation with multi-organ dysfunction, probably as a complication of severe pre-eclampsia and expired on 5th post-operative day.

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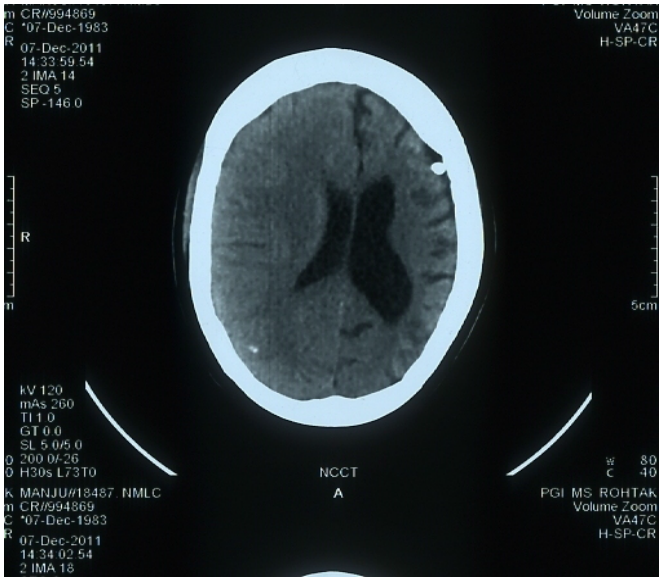

Figure 1: NCCT Picture.

Discussion

For planning the management of recurrent seizures, first step is to identify the cause for seizures. Neuroimaging has proven to be the main tool in investigation of seizure disorder.

Dyke-Davidoff-Masson Syndrome (DDMS) is one amongst the syndromes associated with refractory epilepsy. Dyke-Davidoff and Masson described the plain skull radiographical features of Dyke-Davidoff-Masson Syndrome (DDMS) in 1933. This was described in a series of 9 patients with hemiparesis, seizures, facial asymmetry and mental retardation. The radiographical features of the skull were asymmetry, ipsilateral osseous hypertrophy of the calvarium and hyper-pneumatization of the sinuses [5]. The disease is classified into infantile (congenital) and acquired type. In the infantile type, the various causes propounded are neonatal or gestational vascular occlusion involving the middle cerebral vascular territory, unilateral cerebral arterial circulation anomalies, coarctation of the mid-aortic arch, mesencephalon hypoplasia and wallerian degeneration. These patients become symptomatic in perinatal period or infancy. The acquired type, results from trauma, infection, ischaemia and haemorrhage. The age of presentation depends on time of insult and the characteristic changes may be seen only in adolescence [6].

The mechanism of cerebral atrophy is still unclear, but it is hypothesized that ischemic episodes from a variety of different causes reduce the production of brain derived neurotrophic factors, which in turn lead to cerebral atrophy [7]. 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, the increased width of diploic space and the elevations of the greater wing of sphenoid and the petrous ridge on the affected side [8]. Both the sexes and either 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 [9].

Narain et al described the case of DDMS in an 18 month-old girl who presented with right sided focal seizures, hemiparesis of same side and delayed milestones. The findings of dilated cortical sulci and widening of ipsilateral diploi reflecting a late onset of brain insult probably of vascular origin involving left middle cerebral artery [9].

Lee et al. described a case of a 17 month-old male child who presented characteristics of DDMS with left focal clonic or tonic-clonic seizures accompanied by left hemiparesis and developmental delay. Brain MRIs demonstrated progressive atrophy of the right cerebral hemisphere with dilatation of the lateral ventricle, expansion of the ipsilateral frontal sinus with calvarial thickening, and elevation of the petrous pyramid and orbital roof. Brain Single-photon emission computed tomography (SPECT) showed a decreased volume of the right hemisphere with reduced blood flow [10].

Amann et al. report the first case of left cerebral hemiatrophy and a late onset of treatment-resistant schizoaffective disorder after a stressful life event. Magnetic resonance imaging (MRI) revealed left cerebral hemiatrophy with loss of white matter and gliosis in frontal, temporal, and in posterior areas. Pyramidal degeneration and a left mesencephalic hemiatrophy with a slimming of the corpus callosum were also noted. A slight atrophy at the right hemisphere and cerebellum was observed [2].

The condition needs to be differentiated from Basal ganglia germinoma, Sturge Weber syndrome, Linear nevus syndrome, Fishman syndrome, Silver-Russell syndrome and Rasmussen encephalitis [11,12].

The manifestations of DDMS may be as subtle as to be overlooked on plain radiographs; however, CT is the diagnostic modality of choice [3]. *Lee and Amann* had diagnosed DDMS on MRI [2,10].

The treatment of DDMS with multiple anti-epileptics is the best option. If the seizures are refractory, hemi-spherectomy is the best treatment option [4].

On searching the literature, no case of DDMS in pregnancy could be found and our case appears to be the first one. Generalized seizures during pregnancy can lead to increased maternal trauma which can result in intra-cranial haemorrhage in foetus and if maternal abdomen is involved, a theoretical risk of abruption exists, possibly leading to foetal hypoxia or death. Furthermore, the risk of maternal aspiration can cause maternal hypoxia, which can also lead to fetal hypoxia. Repeated convulsions during pregnancy can significantly increase the maternal and foetal morbidity and mortality. In the present case, the foetus died in utero but the likely cause of intrauterine death appears to be abruptio placentae due to severe pre-eclampsia. Apart from seizures the precise effect of DDMS on foetus is not documented in literature as this is the first reported case of DDMS in pregnancy.

In our case, exact diagnosis could not be made in ante-natal period due to the incomplete history given by patient and attendants as the tendency in developing countries is to hide the facts about chronic disease. CT scan was delayed due to limitation of radiation exposure in pregnancy though the period of teratogenesis was over but still there does exist a risk of carcinogenesis. At the same time the patient responded to anticonvulsants initially and the facility of MRI was not available at that time in the hospital. It could have been diagnosed and managed earlier but due to low socioeconomic status and unawareness, the disease could not be diagnosed till adulthood. No definitive management (hemi-spherectomy) could be done in post-operative period as the general condition of the patient was poor.

Conclusion

DDMS can be diagnosed by the assessment of patient by complete clinical history and examination along with radiological features on CT/MRI. In patients of recurrent seizures, CT/MRI should be considered as early as possible.

Conflict of interests

Authors have no conflict of interests to disclose.

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