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Case Report

Adult Presentation of Dyke Davidoff Masson Syndrome

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Abstract

Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological disorder that results from brain injury in intrauterine or the first years of life. Clinical manifestation are usually hemiplegia or hemiparesis, seizure, facial asymmetry, learning difficulties or mental retardation. Rarely, sensory symptoms or psychiatric disorders may be seen. Brain imaging finding include prominent cortical sulci, dilated lateral ventricle, cerebral hemiatrophy, hyper pneumatization of the frontal sinus, and compensatory hypertrophy of the skull. We describe a female patient with no previous childhood history of brain damage, who presented with generalized tonic-clonic seizure, right-sided body weakness, neuroimaging findings of cerebral hemiatrophy, left lateral ventricle dilatation and homolateral frontal sinus hyperpneumatization.

Keywords: *Dyke Davidoff Masson Syndrome; Epilepsy; Cerebral Hemiatrophy; Magnetic Resonance Imaging*

Abbreviations

DDMS: Dyke Davidoff Masson Syndrome; MRI: Magnetic Resonance Imaging

Introduction

Dyke Davidoff Masson syndrome (DDMS) is a rare syndrome, sometimes under-diagnosed. It refers to cerebral hemiatrophy, contralateral hemiparesis, and epilepsy. Most affected patients are among the pediatric population [1,2].

On brain imaging, the findings confirm the diagnosis and allows the elimination of mimikers. The significance of early and correct diagnosis is the prospect of improving the patient's prognosis and improving their quality of life [3].

Here we present a case of a young girl who was put on anticonvulsants for three years without any exploration. It was only when her symptoms worsened that brain imaging was performed. Hence the need for awareness among epilepsy care providers.

Case Presentation

A 27-year-old female patient presented to our hospital for right side body weakness, walking difficulty and frequent convulsion. The symptoms started three years ago, with generalized convulsive seizure without post-critical deficit, she was evaluated at local health

center and diagnosed with epilepsy and started an anticonvulsant treatment. Later, she developed progressive right side hemibody weakness and gait difficulty. Despite the treatment, the seizure was not controlled; so she was referred to our hospital for further management. There was no history of perinatal and antenatal complications, she was born as a full term baby via normal vaginal delivery and she had normal childhood development.

On neurologic examination, she had spastic right side hemiparesis with brisk deep tendon reflex. Otherwise normal cognition, cranial nerves and sensory system. No signs of inflammation or neurocutaneous lesion were seen in the skin and all routine blood tests were normal.

Brain CT scan was performed (Figure 1), showing the atrophy of the left cerebral hemisphere with compensatory hyper pneumatization of the homolateral frontal sinus, a complementary MRI (Figure 2) confirmed the CT scan diagnosis, showing the atrophy of the left cerebral white matter with thinning of the cortical gyri, dilatation of left lateral ventricle, and normal right cerebral hemisphere.

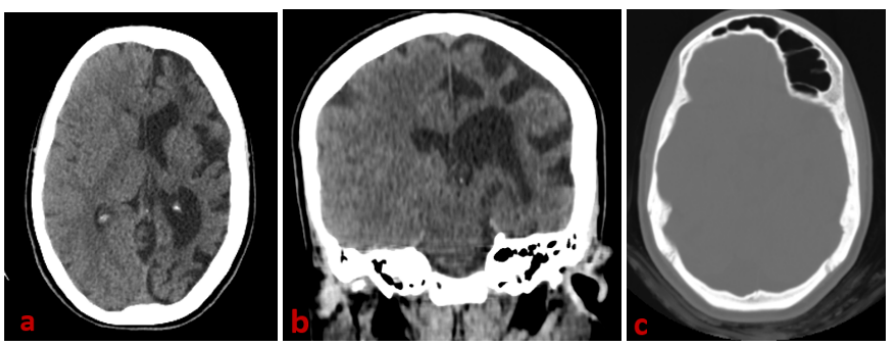

Figure 1: *Axial section (a) with coronal reconstruction (b) showing the atrophy of the left cerebral hemisphere and the dilatation of homolateral ventricle with the compensatory hyper pneumatization of the left frontal sinus (c).*

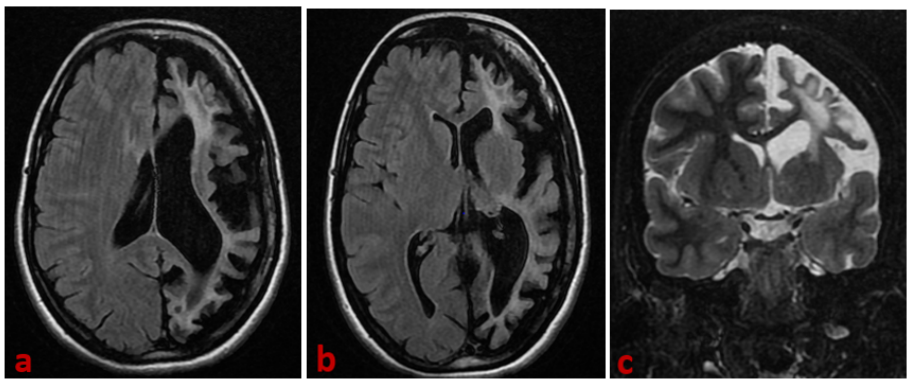

Figure 2: *Axial FLAIR images (a, b) showing the left cerebral hemi atrophy and Coronal T2 image (c) showing the atrophy of the white matter with thinning of the cortical gyri, notice the normal right cerebral hemisphere.*

Discussion

DDMS was first reported by Dyke, Davidoff, and Masson in 1933 [4]. However, a French anatomist and pathologist named Jean Cruveilhier (1791 - 1874) provided an early clinical-pathologic description of Dyke-Davidoff-Masson syndrome. Cruveilhier's case was initially published around 1830, more than a century before the clinical and radiologic report of Dyke and colleagues (1933) based on a series of patients studied with pneumoencephalography [5].

DDMS probably result from intrauterine conditions or if brain damage occurred in the first 3 years of life [4]. There are congenital and acquired etiologies. Congenital causes include congenital malformation, infection and vascular injury (occlusion in the middle cerebral artery), intra uterine anoxia or hypoxia, and intracranial haemorrhage in the perinatal period. Acquired causes are usually haemorrhagic or ischemic vascular diseases in the postnatal period. In addition, damage can be caused by trauma, tumors, prolonged febrile seizure, radiation and infection. The emergence of clinical symptoms in acquired DDMS can begin in childhood or adolescence, rarely in adulthood, depending on the time and quality of the etiological factors. A study of M Atalar., *et al*. of 19 cases, showed that the possible etiology was congenital in 12 patients, and there were acquired causes in 7 cases [6,7]. Some studies have suggested that the left hemisphere is more susceptible to cortical damage since brain blood flow in children between 1 and 3 years of age shows dominance in the right hemisphere [4,8]. Due to the high flow of blood in the right hemisphere, the left hemisphere is more susceptible to cerebrovascular events, especially in infants [8].

Clinical manifestations depend upon the level of damage and consist of contralateral hemiplegia, hemiparesis, seizure, facial asymmetry, language disorders, learning difficulties, and mental retardation. Sensory symptoms and psychiatric disorders such as schizophrenia are occasionally seen [9]. Seizures may be the first manifestation or start months or even years after onset of hemiparesis and mental retardation, although some patients never have any seizures at all [7,9].

Radiological characteristics of DDMS on CT or MRI include unilateral diffuse or focal cerebral parenchyma loss due to encephalomalacia, sulcal enlargement, and ventriculomegaly, in addition, in the same side, compensatory changes are observed, such as calvarial thickening, hyperpneumatization of the paranasal sinuses, and elevation of the temporal bone, and more rarely, hypoplasia of thalamus, lentiform nucleus, caudate nucleus and mesencephalon may be present. The contralateral hemisphere is normal [1,3,4,7]. Radiological findings may vary according to the duration and extent of cerebral injury [4,7]. It has been reported that in order to have calvarial changes, brain damage must have occurred in the intrauterine period or in the first three years of life before calvarial maturation is complete. Therefore, when calvarial damage with cerebral haemiatrophy is observed in patients with DDMS of unknown etiology, it should be assumed that brain damage occurred during the first 3 years of life and went unnoticed [7].

Differential diagnoses of DDMS include Sturge Weber syndrome, Rasmussen encephalitis, basal ganglia germinoma, linear nevus syndrome, Silver Russell syndrome, Fishman syndrome, and hemimegaloencephaly [7,10]. However, a careful clinical examination and neuroimaging will differentiate most of these conditions [10]. Facial cutaneous vascular malformations, seizures, glaucoma, mental retardation, and recurrent stroke-like episodes are clinical manifestations of Sturge–Weber syndrome. Intracranial vascular anomalies and leptomeningeal angiomatosis with stasis may lead to ischemia and result in calcification (causing the intracranial tram track sign on CT) along with laminar cortical necrosis and atrophy [10,11]. There are conflicting ideas about the association between Sturge–Weber syndrome and DDMS. While most authors [9,10] mention Sturge-Weber syndrome in the differential diagnosis of DDMS, others [12,13] have reported that Sturge-Weber syndrome may also lead to DDMS. Rasmussen encephalitis is a chronic progressive inflammatory disorder which manifests during childhood between 6 and 8 years of age with similar imaging and clinical findings of hemispheric atrophy, intractable focal epilepsy and cognitive defects. Calvarial changes are not observed in these patients however [7,10]. A rare brain tumor, basal ganglia germinoma, frequently presents with progressive hemiparesis, cerebral hemiatrophy and calvarial changes. Differentiating imaging findings are cystic areas, focal hemorrhages, and mild surrounding edema [10]. Linear nevus syndrome typically involves facial nevus,

recurrent seizures, mental retardation, and unilateral ventricular dilatation similar to cerebral hemiatrophy [7]. Silver–Russell syndrome has a distinct facial phenotype (triangular face, small pointed chin, broad forehead, and thin wide mouth), along with poor growth and delayed bone age, clinodactyly, hemihypertrophy with normal head circumference, and normal intelligence [7]. Fishman syndrome (encephalocranio cutaneous lipomatosis), a rare neurocutaneous syndrome, involves unilateral cranial lipoma with lipodermoid of the eye and is usually accompanied with seizures. Neuroimaging of the condition reveals a calcified cortex and hemiatrophy [10,14].

Conclusion

DDMS is a rare syndrome with wide clinical and radiological spectra that can be variably symptomatic at different stages of life. Although it is diagnosed with epileptic seizure, hemiparesis, or mental retardation in childhood, it can also be diagnosed symptomatically (mild to severe) or asymptomatically in adulthood with radiological findings, as in our case. Early diagnosis provides appropriate treatment of seizures, physiotherapy for hemiparesia, and special education programs for mental retardation. If the diagnosis is delayed, the success of the supportive treatment will be lower than early time.

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