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Radiological Findings of Dyke-Davidoff-Masson Syndrome in An Adult Patient: Case Report

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Abstract

Dyke Davidoff-Masson syndrome (DDMS) is a rare disease characterized by cerebral hemiatrophy due to brain insult in fetal life. Clinical symptoms include seizures, facial asymmetry, hemiplegia or hemiparesis, and mental retardation. Radiological modalities illustrate unilateral loss of cerebral paranchyma and ipsilateral bone changes due to hemiatrophy. The syndrome had been described mainly in children or adolescents, besides a few cases were reported in adults.

We present computed tomography and magnetic resonance imaging findings of DDMS in a 47- year-old woman with status epilepticus.

Key words: Computed tomography; Dyke-Davidoff-Masson syndrome; Magnetic resonance imaging

Case Report

A 47-year-old female was admitted to our emergency department with right sided focal status epilepticus. She had a history of recurrent seizures since chilhood without a definitive diagnosis. She had no detectable mental retardation. Also, physical examination revealed no facial asymmetry. Her laboratory tests were normal. Neurological examination revealed right-sided spastic hemiparesis and hemiatrophy. Computed tomography (CT) scan of head revealed left sided hemiatrophy with dilatation of the left lateral ventricle, widening of ipsilateral sulci with ipsilateral calvarial thickening and hyperpneumatization of the frontal and sphenoid sinuses and mastoid air cells (Figure 1). Magnetic resonance imaging (MRI) of brain demonstrated no signal abnormalities due to gliosis and confirmed bone changes described at prior CT (Figure 2). According to radiological findings, the patient was diagnosed as Dyke-Davidoff-Masson Syndrome.

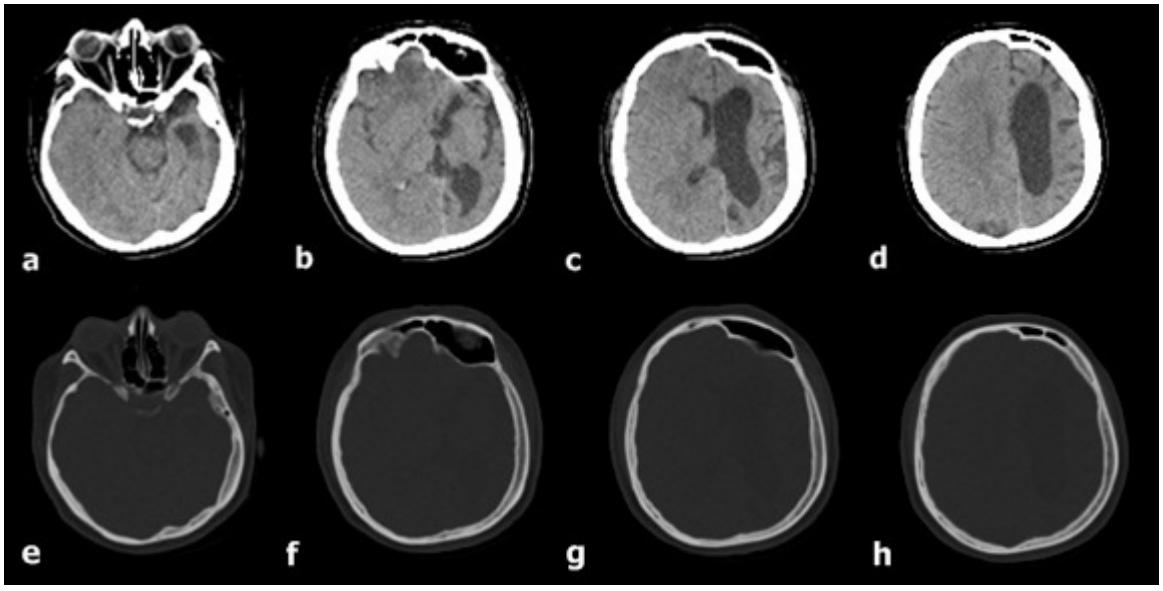

Figure 1: Unenhanced brain CT images demonstrate left sided hemiatrophy with dilatation of the left lateral ventricle **(a-d),** widening of ipsilateral sulci with ipsilateral calvarial thickening and hyperpneumatization of the frontal and sphenoid sinuses and mastoid air cells **(e-h)**

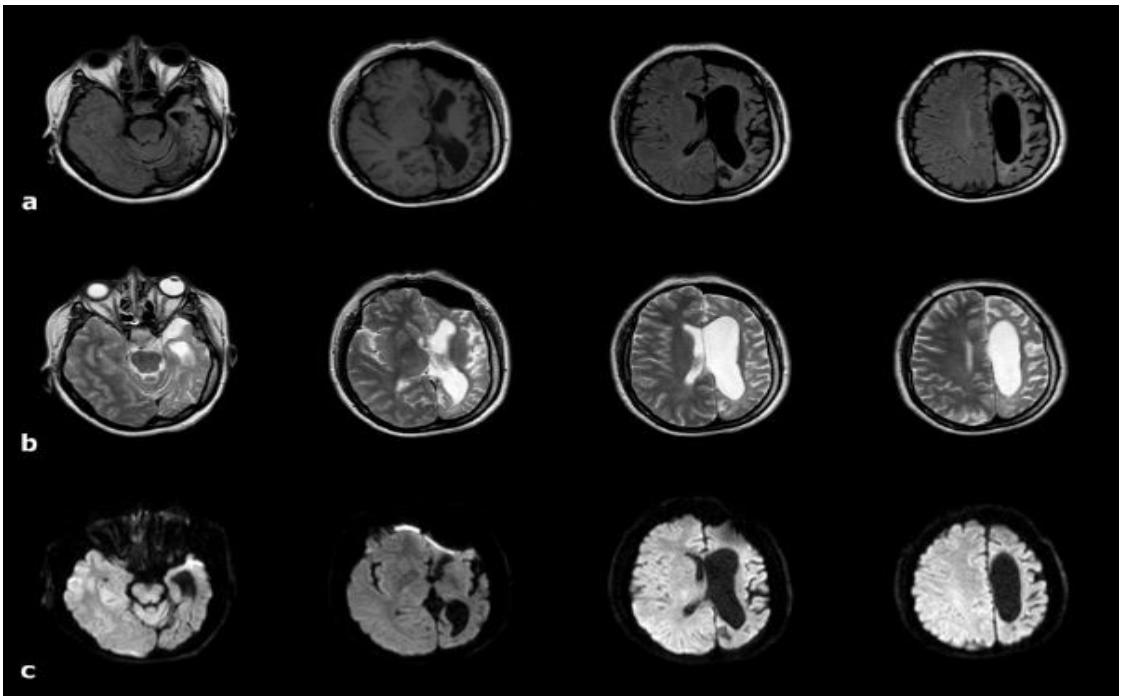

Figure 2: Axial FLAIR **(a)** T2 weighted images **(b)** and diffusion weighted images **(c)** confirme CT findings and demonstrate no signal abnormalities due to gliosis

Discussion

Dyke-Davidoff-Masson syndrome (DDMS) or cerebral hemiatrophy is a rare disease characterized by cerebral hemiatrophy due to brain insult in fetal life [1]. Clinical symptoms include seizures, facial asymmetry, hemiplegia or hemiparesis, and mental retardation. The ethiological factors of DDMS are classified in two; congenital or acquired. In the congenital type, the reason of the damage is vasculary and usually occures in the fetal life. The ethiological factors of acquired type that usually occur during in the perinatal life or later are trauma, infection, ischemia, hemorrahage and vascular disorders [2,3].The clinical features are variable and depend on the extent of brain injury and consists of seizures, facial asymmetry, contralateral hemiplegia or hemiparesis, mental retardation, and speech or language disorders [1,3]. Male sex and left hemisphere involvement are more commonly reported in the literature [4]. In this paper, we report a female patient with left sided hemiatrophy. Although, it is usually diagnosed in the early life or adolescense, a few adult cases are documented [1].

Computed tomography and magnetic resonance imaging demonstrate classical findings of this syndrome such as unilateral cerebral hemiatrophy with ipsilateral compensatory changes of skull and sinuses [3,5-7]. These compensatory bone changes due to relative vacuum effect of the cerebral hemiatrophy are usually seen in the congenital type [3]. MRI is also a valuable radiological modality to reveal associated parenchymal changes and signal abnormalities due to ethiological factors and is helpful in differentiating between congenital and acquired types [6]. Shen et al. [8] have established a three-category model of cerebral hemiatrophy according to MR imaging findings. 1) diffuse cortical and subcortical atrophy; 2) diffuse cortical atrophy associated with porencephalic cysts; and 3) gliosis due to previous infaction in the middle cerebral artery territory. Our patient did not have porencephalic cyst or signal abnormalities due to gliosis. Hence, we considered category 1 according to our patient's MRI findings [9].

In the differential diagnosis, there are other conditions such as hemimegalencephaly, Sturge-Weber syndrome, and Rasmussen encephalitis that are associated with cerebral hemiatrophy [2]. Sturge-Weber syndrome can also be an association and is characterized by facial hemangiomatosis. Rasmussen encephalitis tends not to have calvarial changes [5]. A proper history, physical examination and radiological modalities are helpful in the differential diagnosis.

Conclusion

In conclusion, DDMS is a rare disease that was documented especially in the children and adolescents. Contrary, in the present study, we described an adult patient with focal epileptic seizure due to DDMS, and its classical radiological findings. Radiological modalities especially MRI are very useful for the diagnosis and demostrating associated abnormalities. Hence, the radiologists should have the knowledge of the classical findings of DDMS to avoid misdiagnosis.

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