peer reviewed **CASE REPORT**

A case report on a Dyke Davidoff Masson syndrome with right hemisphere involvement

Chetty, Seshree *BTech: D (DUT)*

Specialist Radiographer, East Surrey Hospital, UK

Abstract

Dyke Davidoff Masson syndrome (DDMS) is a rare condition characterised by seizures, contralateral hemiparesis, mental retardation, behavioural and learning disabilities. It is characterised according to congenital and acquired groups. Either of the hemispheres can be affected in males and females. It more prevalent in males, and left hemisphere involvement is more frequent. This case report describes Dyke Davidoff Masson syndrome with right hemisphere involvement in a female patient. The patient's clinical history, radiological findings, the epidemiology and treatment for DDMS are discussed.

Keywords

Seizures, calvarial thickening, cerebral hemi-atrophy.

Case report

A female in her twenties presented at a Durban state hospital. She was a known epileptic. Following an epileptic episode she fell down a flight of stairs and sustained a mild head injury. There was bruising and swelling over her left temporal region. She had long standing left hemiparesis. Her clinical history was provided by a relative who had accompanied her, However, it was not comprehensive as the patient did not reside with her relative.

An unenhanced computed tomography (CT) scan of the brain was performed to exclude skull fractures or bleed since she had a Glasgow coma scale reading of 13/15. The scan covered the skull base to vertex. The CT findings were asymmetry of the ventricular system with associated right lateral ventricular enlargement (see Figure 1). There was the presence of cerebral atrophy with prominence of the sulcal pattern to the right (Figure 2). There was enlargement of the right paranasal sinuses (Figure 3), and thickening of the calvarium on the right (Figure 4). A left sided subgaleal hematoma was noted with marked soft tissue swelling.

The features were suggestive of hemiatrophy; also known as Dyke Davidoff Masson syndrome (DDMS). Surgical intervention was not considered. She was prescribed drugs to control her seizures, and was placed on twenty-four hour neurology observations.

Discussion

Dyke Davidoff Masson syndrome (DDMS) is characterised by cerebral hemi-atrophy, seizures, hemiplegia on the contralateral side, facial asymmetry, and mental retardation which is most likely thought to be caused by a decrease in vascular supply.[1, 2] There are many causes of DDMS , and it is categorised according to congenital and acquired groups.[3] This condition affects both genders and both hemispheres can be affected.[1, 3] However, it occurs most frequently in males with involvement of the left hemisphere.[1, 3] It is caused by developmental hypoplasia or acquired atrophy of one cerebral hemisphere, which results in a wide range of clinco-pathological issues.[4] DDMS is characterised by cerebral hemi-atrophy and or cephalic disorder with ipsilateral facial hypoplasia and contralateral skeletal deformities.[5] Epilepsy is one of the main features of this syndrome; it is also known as a rare epilepsy syndrome, which is defined by a typical range of imaging and clinical features.[4, 6] This patient was an epileptic and had left sided hemiparesis.

DDMS is a type of focal cerebral atrophy that involves one cerebral hemisphere. It includes various groups of clinical and pathological characteristics with decreased growth of the affected hemisphere.[2] It affects males and females, and involves either hemispheres of the brain. However, left hemisphere involvement in males is more common.[2, 3] There are reported cases of right hemisphere involvement in females.[7]

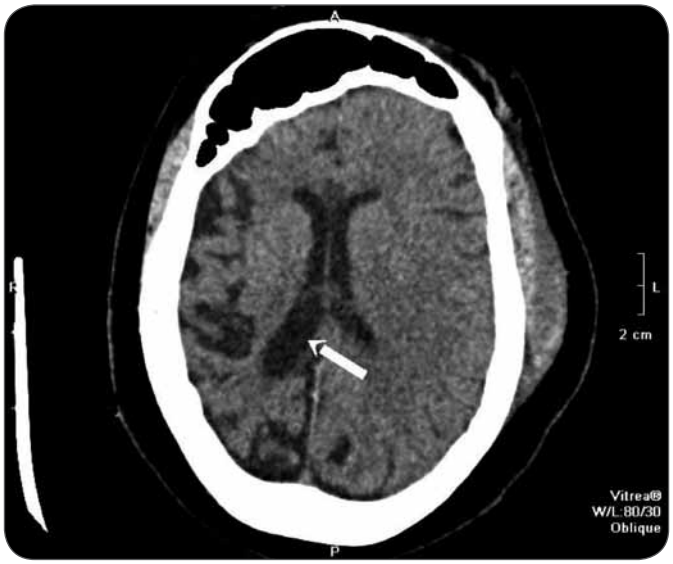

Figure 1. Axial unenhanced CT brain scan, showing asymmetry of the ventricular system and enlargement of the right ventricle (arrow).

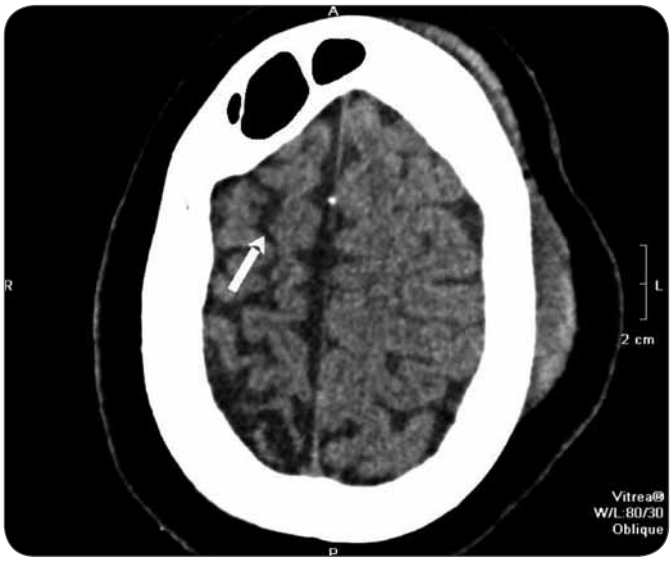

Figure 2. Axial unenhanced CT brain scan, showing cerebral hemi-atrophy of the right hemisphere with enlarged sulci (arrow) as compared to the left hemisphere.

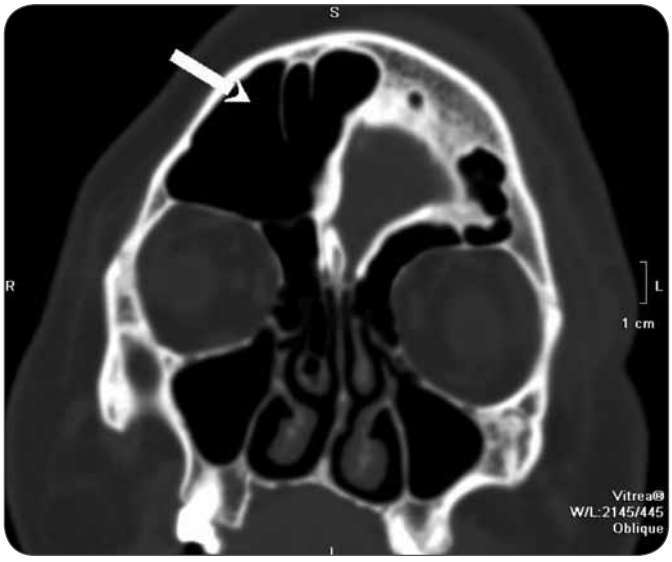

Figure 3. Coronal unenhanced CT scan of the sinuses, showing enlargement of the right paranasal sinuses (arrow) compared to the left paranasal sinuses.

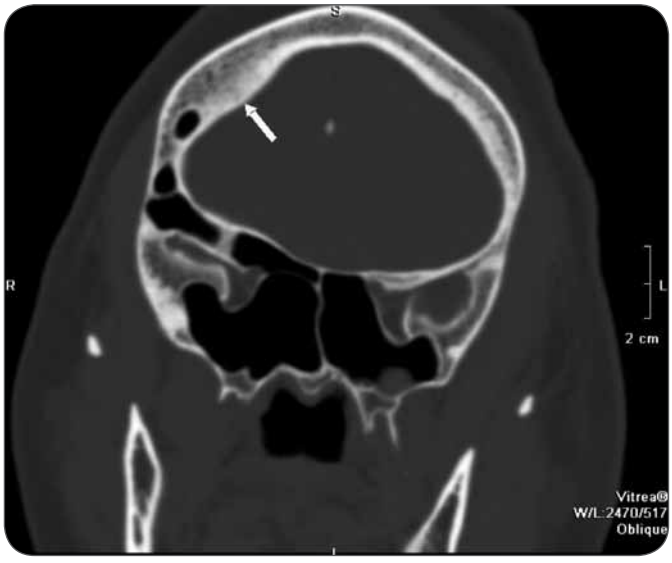

Figure 4. Coronal unenhanced CT brain scan showing thickened calvarium on the right (arrow).

This condition is categorised according to congenital and acquired groups. The former is triggered mostly due to vascular obstructions or deformities in-utero or in the neo-natal period.[3, 4] The latter is caused through infection, trauma, ischemia and haemorrhage.[1, 4] The difference between these two types is that in congenital type there is shift of midline structures towards the side of the disease and the sulcal prominence replacing the gliotic tissue is absent. This feature differentiates it from cerebral hemi-atrophy that occurs later in life.[3] Its clinical features include lack of facial symmetry, seizures, spastic hemiplegia and contralateral hemiparesis and mental retardation.[2, 6] Clinical presentation also shows deviation according to the degree of brain injury.[4]

In the case of the acquired type, age of presentation is determined by the time of neurologic trauma. Clinical findings may be visible only in the teenage years.[1] Clinical features differ depending on the degree of brain injury.[3]

The most significant radiological features of this syndrome include a unilateral loss of cerebral volume, calvarial thickening, enlargement of the paranasal sinuses and petromastoid air cells, a raise in the sphenoid wing and petrous ridge and ipsilateral osseous hypertrophy.[3, 4, 6] The CT scan for this patient showed these radiological features.

CT, positron emission tomography (PET), and magnetic resonance (MRI) imaging, can be used to see the extent of cerebral parenchymal involvement.[3, 6] The glucose metabolic differences seen by PET are in keeping with the atrophic changes visible on MRI.[6]

The treatment is symptomatic, and includes management of convulsions, hemiplegia, hemiparesis and learning difficulties. Prospects are better if hemiparesis occurs after the age of two years, and in the lack of prolonged or recurrent seizures. Children with intractable disabling hemiplegia are the likely candidates for hemisphrectomy with a success rate of 85 percent in cautiously selected cases.[3]

This condition needs to be separated from basal ganglia germinoma, Sturge-Weber syndrome, Linear Nevus syndrome, Fishman syndrome, Silver-Russel syndrome, and Rasmussen encephalitis.[3] An accurate history, complete clinical examination and radiological findings provide the precise diagnosis.[1]

Conclusion

This female patient was diagnosed with DDMS which involved the right hemisphere of the brain, even though it is noted that this syndrome predominately appears in males with left hemisphere involvement. CT played an important role in the diagnosis and management of this patient. This patient was managed with medication.

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