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Dyke-Davidoff-Masson Syndrome Revealed by Refractory Epilepsy in a Young Adult: A Case Report

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AUTHORS AND AFFILIATION

Mehdi Salmane[1], Abdallah Said[1], Alia Yassine Kassab[1], Lina Lasri[1], Firdaous Touarsa[1], Meryem Fikri[1]

[1] Department of Neuroradiology, Specialties Hospital, Rabat

Corresponding author: Mehdi Salmane.

ABSTRACT

Dyke-Davidoff-Masson syndrome (DDMS) is a rare neurological condition characterized by cerebral hemiatrophy associated with compensatory osseous hypertrophy. We report the case of a 24-year-old man presenting with refractory focal seizures and progressive left-sided weakness since childhood. Brain MRI demonstrated marked right cerebral hemiatrophy with ipsilateral ventricular dilatation, calvarial thickening, and hyperpneumatization of the frontal sinus, consistent with Dyke-Davidoff-Masson syndrome. This case highlights the characteristic imaging features of this rare entity and the importance of MRI in establishing the diagnosis.

KEYWORDS :

Dyke-Davidoff-Masson syndrome, Cerebral hemiatrophy, Refractory epilepsy, Hemiparesis.

MAIN ARTICLE

INTRODUCTION

Dyke-Davidoff-Masson syndrome (DDMS) is a rare condition resulting from cerebral injury occurring during fetal life or early childhood, leading to cerebral hemiatrophy and compensatory cranial changes. The syndrome classically presents with seizures, contralateral hemiparesis, facial asymmetry, and cognitive impairment.

MRI plays a central role in diagnosis by demonstrating both parenchymal and osseous abnormalities [1,2].

CLINICAL PRESENTATION

A 24-year-old right-handed man was admitted for worsening refractory focal seizures evolving since the age of 5 years. The patient also reported progressive weakness of the left upper and lower limbs associated with mild learning difficulties.

Neurological examination demonstrated:

- Mild left spastic hemiparesis
- Facial asymmetry
- Brisk osteotendinous reflexes on the left side

No history of recent trauma or infection was noted. Family history was unremarkable.

MRI Findings

Brain MRI demonstrated:

- Marked right cerebral hemiatrophy predominantly involving the frontal and parietal lobes
- Ex vacuo dilatation of the right lateral ventricle
- Prominence of cortical sulci on the affected side
- Ipsilateral thickening of the cranial vault
- Hyperpneumatization of the right frontal sinus

No abnormal diffusion restriction or contrast enhancement was identified.

These findings were highly suggestive of Dyke-Davidoff-Masson syndrome.

DISCUSSION

DDMS is attributed to prenatal or early postnatal cerebral insult caused by ischemia, trauma, infection, or vascular malformations. The timing of the cerebral injury influences the degree of compensatory osseous hypertrophy.

Clinical Presentation

Patients typically present with:

- Refractory seizures
- Contralateral hemiparesis
- Developmental delay
- Facial asymmetry

In this case, epilepsy was the predominant symptom leading to imaging evaluation [2].

Radiological Features [3]

Characteristic MRI findings include:

- Unilateral cerebral hemiatrophy
- Ipsilateral ventricular enlargement
- Calvarial thickening
- Enlarged paranasal sinuses and mastoid cells
- Gliosis and encephalomalacia

These compensatory osseous changes usually develop when cerebral injury occurs before 3 years of age.

Differential Diagnosis

Differential diagnoses include:

- Rasmussen encephalitis
- Sturge-Weber syndrome
- Hemiconvulsion-hemiplegia-epilepsy syndrome
- Post-traumatic cerebral atrophy

MRI findings are generally characteristic and allow confident diagnosis [3, 4].

Management

Treatment is mainly symptomatic and includes:

- Antiepileptic therapy
- Physiotherapy
- Cognitive rehabilitation

In severe refractory epilepsy, hemispherectomy may be considered in selected patients [3,5].

CONCLUSION

This case illustrates a classic presentation of Dyke-Davidoff-Masson syndrome in a young adult with refractory epilepsy and chronic hemiparesis. MRI demonstrated typical cerebral and calvarial abnormalities, allowing straightforward diagnosis. Recognition of these characteristic imaging findings is essential for appropriate neurological management.

FIGURES

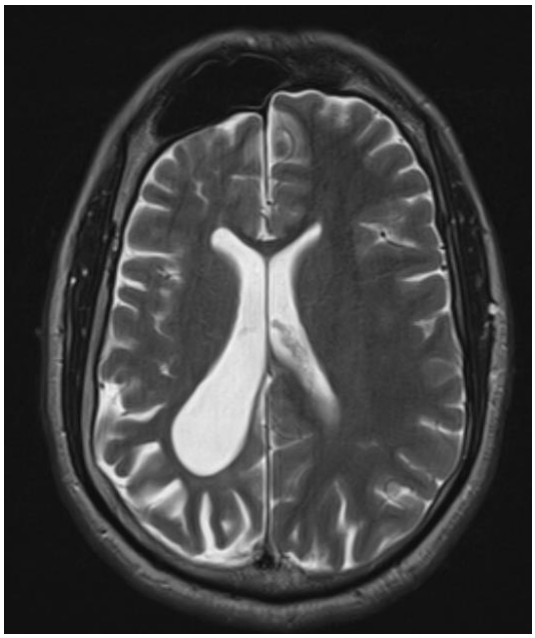

Figure 1: Axial T2-weighted MRI image demonstrating marked right cerebral hemiatrophy with ex vacuo dilatation of the ipsilateral lateral ventricle.

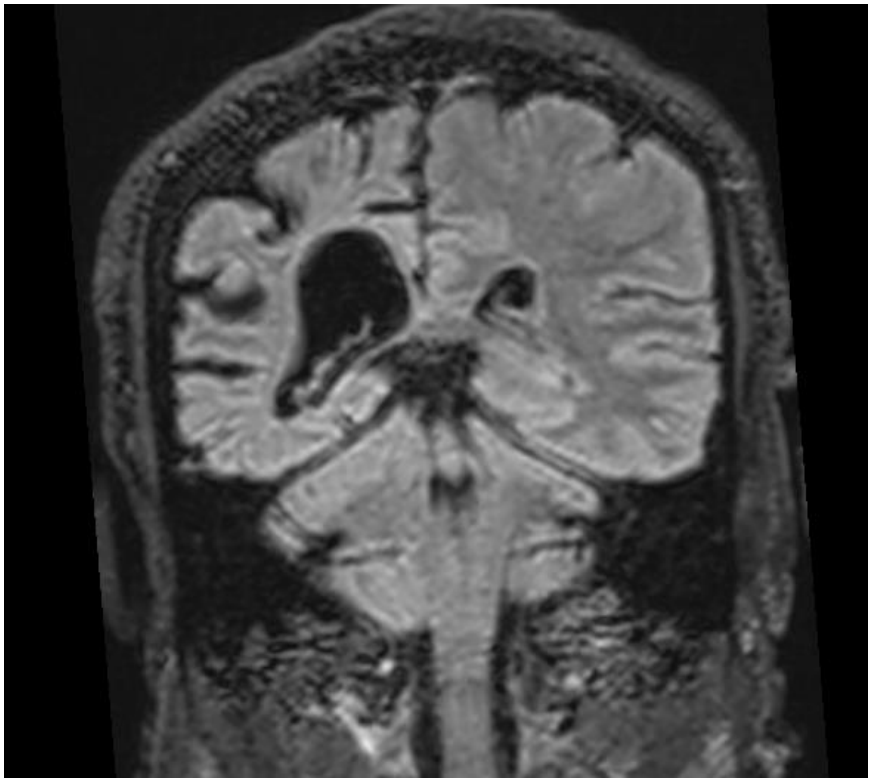

Figure 2: Coronal FLAIR image showing cortical volume loss and prominent sulci within the right cerebral hemisphere.

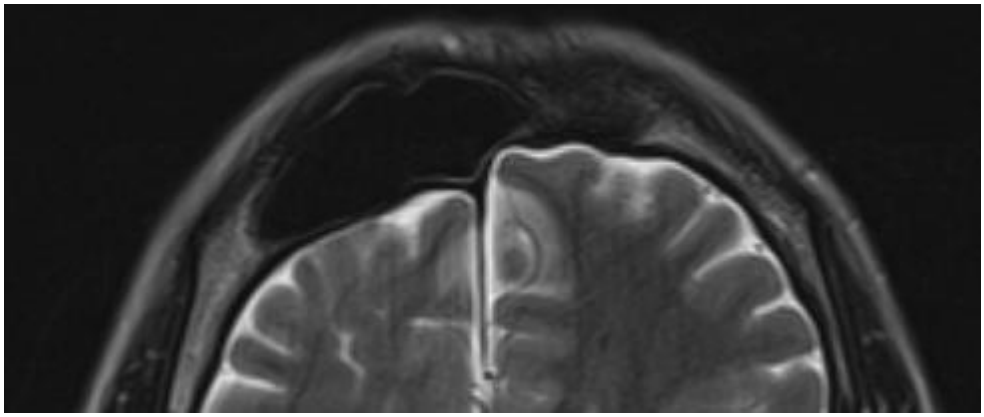

Figure 3: Axial MRI image illustrating compensatory hyperpneumatization of the frontal sinus.

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