

Neurosciences Quiz

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A patient with left hemiparesis, seizure, and development delay on admission

Clinical Presentation

A 21-month-old boy was admitted to our hospital with the complaint of seizures since 12 months of age. He also had left hemiparesis since 8 months of age. He reached developmental milestones slowly, such as stood up by himself at 19 months of age. On neurological examination, he had left hemiparesis, right hand preference, and facial asymmetry. The rest of the examinations were unremarkable. Cerebral magnetic resonance imaging (MRI) is shown (Figures 1 & 2).

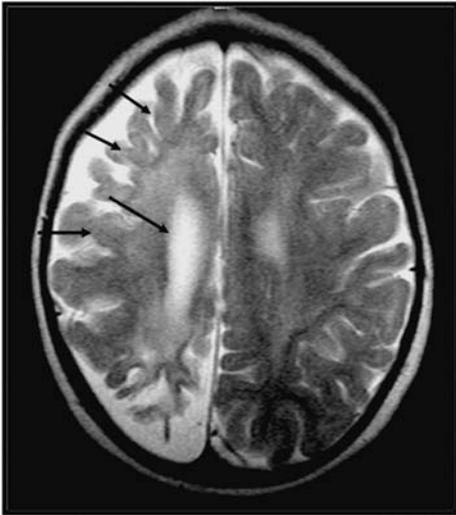


Figure 1 - Axial magnetic resonance images of patient.



Figure 2 - Coronal magnetic resonance images of patient

Questions:

1. What abnormalities are seen in Figure 1?
2. What abnormalities are seen in Figure 2?
3. What is the diagnosis?

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Answers

1. Axial MRI image showed right cerebral hemiatrophy, ipsilateral dilatation of ventricle, and compensatory hypertrophy at the contralateral hemisphere.
2. Coronal MRI image showed right cerebral hemiatrophy, and compensatory hypertrophy at the contralateral hemisphere.
3. Dyke-Davidoff-Masson syndrome (cerebral hemiatrophy). Major signs of this syndrome are cerebral hemiatrophy, contralateral hemiplegia, and convulsions.¹

Discussion

Dyke-Davidoff-Masson syndrome (DDMS) is a rare congenital malformation first reported by Dyke, Davidoff, and Mason in 1933.¹ Cerebral hemiatrophy also known as DDMS, is a condition characterized by seizures, facial asymmetry, contralateral hemiplegia or hemiparesis, and learning difficulties.² Diagnosis is made primarily by clinical features and radiological findings. Radiologic findings are unilateral loss of cerebral volume and ipsilateral calvarial compensatory changes, such as thickening of calvarial bones, hyperpneumatization in paranasal sinuses, and elevation of the temporal bone.³

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References

1. Dyke CG, Davidoff LM, Masson CB. Cerebral hemiatrophy with homolateral hypertrophy of the skull and sinuses. *Surg Gynecol Obstet* 1933; 57: 588-600.
2. Aguiar PH, Liu CW, Leitão H, Issa F, Lepski G, Figueiredo EG, et al. MR and CT imaging in the Dyke-Davidoff-Masson syndrome. Report of three cases and contribution to pathogenesis and differential diagnosis. *Arq Neuropsiquiatr* 1998; 56: 803-807.
3. Zeiss J, Brinker RA. MR imaging of cerebral hemiatrophy. *J Comput Assist Tomogr* 1988; 12: 640-643.

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- * Supplements will be considered for work including proceedings of conferences or subject matter covering an important topic.
- * Material can be in the form of original work or abstracts.
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