



When Asymmetry is the rule only Dyke & Davidoff Can Help An Interesting Case of Cerebral Hemiatrophy (Clinical Image)

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Abstract

Here we present an interesting case of hemiatrophy of the brain resulting from any insult to the growing brain, which leads to corresponding phenotypic manifestations in the body.

Keywords: *Hemiatrophy, Dyke-Davidoff-Mason syndrome.*

Case Details

A 27 year old gentleman was referred from the ophthalmology department in view of dysmorphic facial features. On evaluation he was found to have a hemi facial atrophy, hemiparesis on the left side. There was no other significant past history. On routine imaging studies, he was found to have a grossly asymmetrical brain, with atrophy of all cerebral hemispheres proportionately on the right side along with increased thickness of the skull on the affected right side as shown.

Dyke-Davidoff-Masson syndrome is a clinical syndrome characterized by hemiparesis, facial hemiatrophy, seizures, speech impairment and occasionally mental retardation¹. It is most often seen in the male gender.² It was first described by the three clinicians in 1933 in a series of 9 patients describing in detail their clinical, radiological characteristics³. The MRI features include the presence of cerebral hemiatrophy, calvarial

thickening on the same side, pneumatisation of the sinuses on the same side¹.

The differential diagnosis⁴ in such cases to be considered are:

1. Dyke-Davidoff-Masson syndrome
2. Silver-Russel syndrome
3. Sturge-Webber syndrome
4. Rasmussen's encephalitis

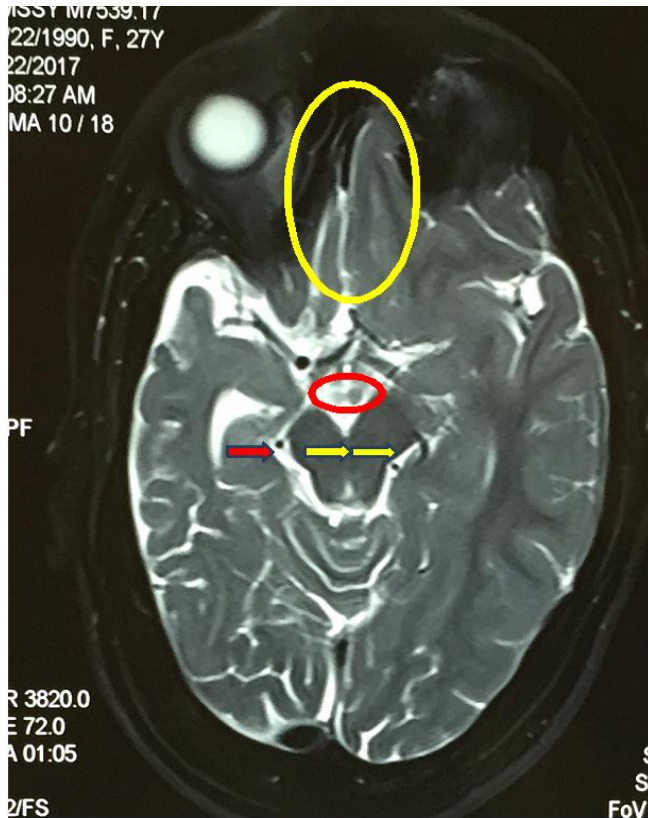


Figure-1: Shows an axial T2 weighted image which shows

1. **Yellow Oval**- Grossly asymmetrical **gyrus rectus**, right atrophied compared to left
2. **Red Oval** – Atrophied **mammillary body** on the left compared to the right
3. **Yellow Arrow** – Gross asymmetry in the sizes of the **cerebral peduncles**
4. **Red Arrow**– Dilated **temporal horn** of lateral ventricle on the right side as compared to the left side.

1. **Yellow Oval**- Sizes of the two hemispheres, along with the sulci-gyri patterns.
2. **Red Arrow**- Dilated Lateral ventricles on the affected right side.
3. **Yellow Brackets**- Thickness of the calvaria more on the right side. (although CT would have been more ideal modality to assess the skull thickness)

References

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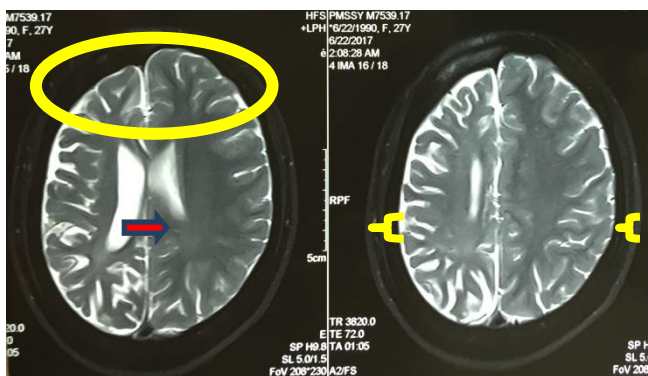


Figure-2: Shows an axial T2-Weighted MRI image which shows the characteristic asymmetry in the: