

## Pictorial CME

### A Girl with Seizure Disorder

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A twelve year old girl born full term to non-consanguineous parents presented with recurrent generalized seizures for last 4 years. She had learning disability, delayed milestones and difficulty in walking. There is no history of birth trauma, antenatal or perinatal complications except delayed cry after birth.

On examination, she had mental retardation, upper motor neuron signs in left upper and lower limbs and mild deviation of mouth to the right. Bilateral carotid pulsations were normal.

Her investigations revealed an abnormal EEG suggestive of generalized

seizure disorder (Fig 1). The MRI showed hemi-atrophy of the right cerebral hemisphere (Fig 2) with ipsilateral dilatation of the lateral ventricle.



Fig 1 — EEG demonstrating generalized seizure disorder

Thickening of the calvarium was noted on the right (Fig 3) along with prominence of right mastoid air cells. (Fig 4) A diagnosis of Dyke Davidoff Masson Syndrome was made.

Childhood Cerebral Hemi-atrophy Syndromes may be congenital or acquired. Acquired causes include trauma, perinatal infections as well as Rasmussen encephalitis<sup>1</sup>, ischemia, neoplasia and radiation injury. Some disorders like Sturge Weber Syndrome<sup>2</sup> neurofibromatosis, Parry Romberg Syndrome<sup>3</sup>, Silver Russel Syndrome may also present with cerebral hemi-atrophy.

The Dyke Davidoff Masson Syndrome, is characterized by mental retardation, epilepsy, hemiparesis and psychiatric manifestations. There is hemi-atrophy of the cerebral hemisphere, ventricular



Fig 2 — Hemiatrophy of right cerebral hemisphere with ipsilateral dilatation of the lateral ventricle

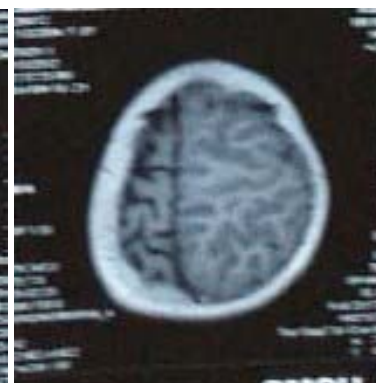


Fig 3 — Thickening of the calvarium of the right side

dilatation with calvarial thickening, along with overpneumatization of mastoid and paranasal sinuses<sup>4</sup>. The calvarial changes signify that the cerebral atrophy happened early in life as  $\frac{3}{4}$ <sup>th</sup> of brain growth occurs before 3 years of age. The acquired causes lack the osseous changes.

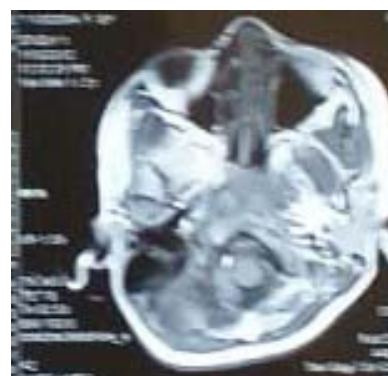


Fig 4 — Prominence of the right mastoid cells

#### REFERENCES

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