Dyke-Davidoff-Masson Syndrome Case 2

Clinical History: An 8-year-old female presented by his parents in pediatric emergency with seizures, right side hemiplegia and mental retardation. She had hypoxic ischemic insult. On examination she had spasticity in right side of body and mental retardation.

Clinical History: An 8-year-old female presented by his parents in pediatric emergency with seizures, right side hemiplegia and mental retardation. She had hypoxic ischemic insult. On examination she had spasticity in right side of body and mental retardation.

Figures 1a and 1b: MRI T2W axial images show left cerebral hemiatrophy, dilatation of ipsilateral lateral ventricle and ipsilateral mid line shift of 7 mm.
**Figure 2:** Saggital T2W image of concerned area.
Figures 3a and 3b: FLAIR axial and coronal images at the level of lateral ventricle show gliotic area with atrophy of left parietal lobe with relative preservation of deep grey nuclei.

Figures 4a and 4b: MRI diffusion weighted and ADC mapping images does not show restricted diffusion in concerned area. Brain MRI was performed which revealed atrophy of left cerebral hemisphere, cerebral peduncle.
There was ipsilateral midline shift and ventricular dilatation along with skull vault thickening and prominent frontal sinus, suggestive of congenital type of cerebral hemiatrophy or Dyke-Davidoff-Masson Syndrome.

**Diagnosis:** Dyke-Davidoff-Masson Syndrome

**MRI findings:** There is presence of left cerebral atrophy involving left frontal, parietal and temporal lobe with dilatation of ipsilateral lateral ventricle and ipsilateral mid line shift of 7 mm towards left side with gliotic area in left parietal lobe with ipsilateral calvarial thickening.

**Discussion:** Patient presented with seizures, facial asymmetry, contralateral hemiparesis and mental retardation. The underlying etiology is cerebral insult that may occur in utero or early in life. Prenatal causes include congenital anomalies, cerebral infarction, vascular malformations and infections.

Perinatal causes are birth trauma, hypoxia and intracranial bleed. Postnatal hemiatrophy can develop secondary to cerebral trauma, tumors, infections and febrile seizures. Infantile (congenital) type of DDMS, in contrast to adult (acquired) DDMS, shows enlargement of calvarium, diploic space and paranasal sinuses. These compensatory cranial changes occur to take up the relative vacuum created by the atrophied cerebral hemisphere. Shen et al. depicted three MR imaging patterns of cerebral hemiatrophy: MR imaging pattern I corresponds to diffuse cortical and subcortical atrophy; pattern II corresponds to diffuse cortical atrophy coupled with porencephalic cysts; and pattern III corresponds to previous infarction with gliosis in the middle cerebral artery (MCA) territory. In our case, pattern III was present. The atrophied cerebral hemisphere will have prominent sulcal spaces if the vascular insult occurs after birth or after end of sulcation. However, if ischemia occurs during embryogenesis when the formation of gyri and sulci is deficient, prominent sulcal spaces will be absent.

Children with medically refractive epilepsy and hemiplegia may be candidates for hemispherectomy, which is helpful in eradicating or significantly reducing seizures in 85 percent of patients. MRI is a valuable method of examination in the analysis of cerebral hemiatrophy as it has the ability to bring to light changes in the cerebral hemispheres as well as highlighting bony structural changes and thus differentiating between congenital and acquired types of DDMS.

**References:**

**Source URL:**
http://www.diagnosticimaging.com/printpdf/dyke-davidoff-masson-syndrome-case-2/page/0/1

**Links:**