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Case Report

Hemimegalencephaly

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Abstract

Hemimegalencephaly is a rare congenital structural malformation of brain due to defective neuronal proliferation, migration and organization, leading to hamartomatous overgrowth of all or part of one hemisphere. There is usually an inverse relation between the size of the affected hemisphere and severity of the malformation. The affected individuals usually present with intractable seizures, contralateral weakness, psychomotor retardation. Treatment is mainly aimed at controlling the seizures.

Keywords: Hemimegalencephaly, unilateral megalencephaly.

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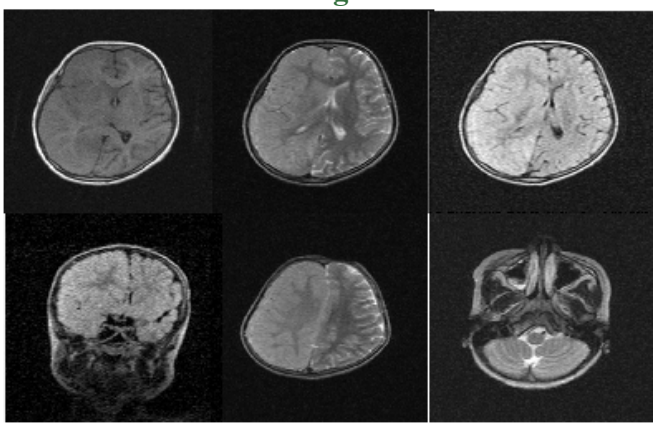
INTRODUCTION

A 8yr old female child presented with left sided hemiparesis, seizure disorder and has a history of developmental delay. Physical examination reveals grade 2 power in left upper and lower limbs. MRI was done.

IMAGING FINDINGS

The right cerebral hemisphere is diffusely dismorphic with broadened gyri and thickened cortical mantle consistent with pachygyria and polymicrogyria. There is poor demarcation of right basal ganglia and thalamus with small corpus callosum. The right lateral ventricle is dymorphic and compressed. There is also subependymal grey matter heteropia. Left cerebral, bilateral cerebellar hemispheres, brainstem are normal. These findings are suggestive of right hemimegalencephaly.

Images



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DISCUSSION

Hemimegalencephaly or unilateral megalencephaly is a congenital disorder in which there is hamartomatousover growth of all or part of a cerebral hemisphere. The affected hemisphere may have focal or diffuse neuronal migration defects, with areas of polymicrogyria, pachygyria, and heterotopia. Hemimegalencephaly is a rare disorder, accounting for only ~2% of childhood epilepsy and was first described by Sims in 1835 after reviewing 253 autopsies. Although the cause is unknown, it is postulated that it occurs due to insults during the second trimester of pregnancy, or as early as the 3rd week of gestation, as a genetically programmed developmental disorder related to cellular lineage and establishment of symmetry. Hemimegalencephalymay also be considered a primary disorder of proliferation wherein the neurons that are unable to form synaptic connections are not eliminated and are thus accumulated.

There are three types of hemimegalencephaly,

Isolated form: The classic and most common type. Sporadic without evidence of cutaneous or systemic involvement.

Systemic form: Associated with partial or total hemigigantism and/or several neurocutaneous syndromes, including epidermal nevus syndrome (sporadic spectrum of disorders that includes linear sebaceous nevus syndrome and the Proteus syndrome)

Total hemimegalencephaly: The least common form, in which there is involvement of the ipsilateral cerebellum and brainstem.

CLINICAL FEATURES

The majority of patients present in infancy with macrosomia, developmental delay and intractable seizures. The older children present with hemiparesis or hemiplegia, mental retardation, seizures and hemianopia.

Imaging features

All the imaging modalities which images brain, can more or less able to identify the key features, but MRI provides best imaging features.

CT FINDINGS

• Enlarged hemisphere and hemicranium.

- The posterior falx often appears displaced across the midline.
- Lateral ventricles are enlarged on affected side, may sometimes be small.
- Shallow sulci with enlarged gyri.
- Abnormal white matter myelination may increase in attenuation, making the contralateral normal white matter unusually hypodense.
- Dystrophic calcifications may be seen in white matter.

MRI FINDINGS

T1: The cortex often appears thickened and 'lumpy-bumpy'.

- Neuronal heteropias are common.
- Ipsilteral ventricle is usually enlarged and deformed, but sometimes may be small.

T2: Areas of lissencephaly, agyria,pachygyria and polymicrogyria are seen in varying proportions.

- Indistinct borders between grey and white matter.
- White matter signal intensity on T2/FLAIR is often heterogenous with cysts and gliosis like hyperintensity.

Differential diagnosis:

1) Enlarged hemisphere
   a) Gliomatosiscerebri
2) Small hemisphere, making the normal hemisphere appear large a) Rasmussen encephalitis
   b) Dyke-Davidoff-Masson syndrome
   c) Sturge-Weber syndrome
3) Other neuronal migration anomalies without overgrowth
   a)polymicrogyria/lissencephaly/agyria/pachygyria

Management:

- The treatment is mainly targetted at controlling seizures, which often can be difficult to control medically.
- In medically refractory seizures, surgery is usually the treatment of choice.
- The surgical procedures that are done are
  (1) Hemispherectomy, with a success rate of ~60% in selected cases.
  (2) Functional hemisperotomy.

REFERENCES

1. Osborn's Brain: Imaging, Pathology, and Anatomy Textbook
2. Broumandi DD, Hayward UM, Benzian JM et-al. Best cases from the AFIP: hemimegalencephaly. Radiographics. 24 (3): 843-8.
3. Abdel Razek AA, Kandell AY, Elsorogy LG et-al. Disorders of cortical formation: MR imaging features. AJNR Am J Neuroradiol. 2009;30 (1): 4-11.

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4. Shorvon SD, Andermann F, Guerrini R. The Causes of Epilepsy, Common and Uncommon Causes in Adults and Children. Cambridge Univ Pr. (2011) ISBN:0521114470.
5. Barkovich AJ, Chuang SH (1990) Unilateralmegalencephaly: correlation of MR imaging and pathologic characteristics. AJNR Am J Neuroradiol 11:523–531
6. Sato N, Yagishita A, Oba H, Miki Y, Nakata Y, Yamashita F *et al*. Hemimegalencephaly: a study of abnormalities occuring outside the involved hemisphere. AJNR Am J Neuroradiol. April 2007;28:678-682.

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