Hemimegalencephaly in a 3-month-old Male Infant: A Case Report from Port Harcourt and Review of the Literature

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ABSTRACT

Introduction of magnetic resonance imaging (MRI) in Port Harcourt, South-South geopolitical region of Nigeria, has led to an increase in the number of diagnosis of congenital cerebral malformations within the locality. Reported cases of hemimegalencephaly from Nigeria and indeed from the West African subregion are very scarce. Here, we report a case seen in Port Harcourt where the diagnosis was made possible by MRI and illustrating the imaging features of this rare condition.

Key words: Congenital cerebral malformation; hemimegalencephaly; magnetic resonance imaging; South-South Nigeria

Case Report

H.E., a 3-month-old male, presented in the magnetic resonance suite of a private radiological center for a cranial scan with a history of tonic-clonic seizure from birth. Pregnancy and delivery had been uneventful and delivery was at term by spontaneous vertex. Head size was normal with no skin lesions seen.

Cranial magnetic resonance imaging (MRI) [Figures 1 and 2] showed enlargement of the left cerebral lobe with thickening of the gray matter. The falx was displaced to the contralateral side with exaggeration of the sulci and gyri. The entire white matter was hyperintense on T2-weighted images. The left cranial vault was also enlarged and the right cerebral hemisphere was reduced in size. The lateral ventricles were enlarged (left worse than the right) with displacement to the right. The third and fourth ventricles were also enlarged.

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A diagnosis of an isolated localized form of hemimegalencephaly (HME) in the left fronto-parietal lobe was made.

Discussion

HME is a rare severe form of cortical developmental malformation characterized by asymmetrical enlargement of all or a part of the cerebral hemisphere. Other features of focal or diffuse neuronal migration include defects such as areas of polymicrogyria (cortical malformation characterized by an excessive number of small, partly fused convolutions [gyri] on the surface of the brain), pachygyria (unusually thick convolutions of the cerebral cortex), and heterotopia (clumps of gray matter located in abnormal areas of the brain).[1] The left cerebral hemisphere is mostly affected in 72.7% of the cases, with the localized form accounting for one quarter of all the cases.[2]

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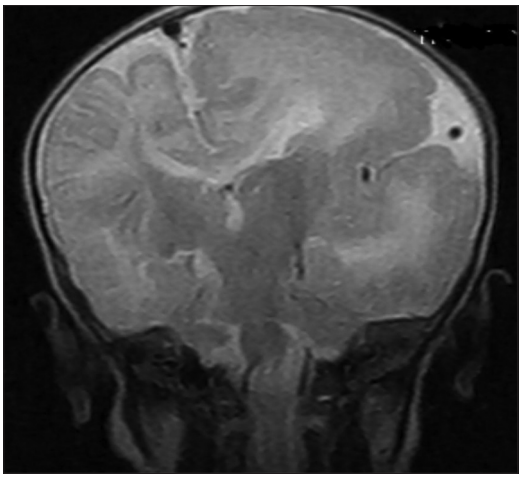

Figure 1: Coronal slice of magnetic resonance imaging of the brain. There is enlargement of the left cerebral hemisphere with dilatation of the ventricles. Thickening of the gray matter and hyperintense white matter

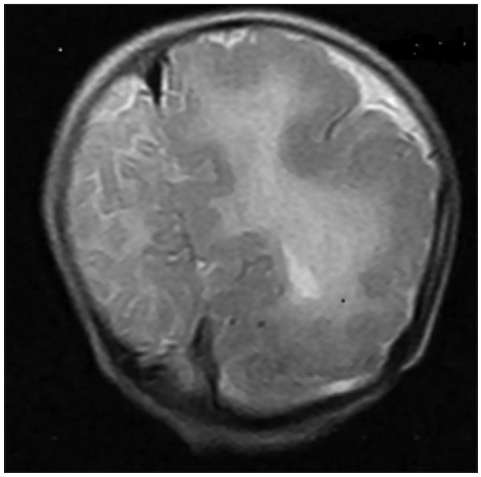

Figure 2: Axial slice of the brain showing enlargement of the left cerebral hemisphere with dilatation of the ventricle and abnormality of the left gray matter intensity

The malformation was first described in 1835 by Sims.[3] Its etiology is unknown; however, it has been thought to be due to abnormal neuronal and cell migration of a single cerebral hemisphere, thus leading to acumination of abnormal neurons. Another school of thought is that it is due to genetic abnormality of neuroepithelial cell lineage and establishment of body symmetry that occurs in the 3rd week of gestation to the 2nd trimester of life.[4] No racial or gender predilection has been noted.[5] It is rare, accounting for only ~0.2% of cases of childhood epilepsy,[5] but makes up 14% of the patients with cortical developmental abnormalities.[6] Our patient presented with a history of tonic-clonic seizures since birth.

HME is classified into three types as follows:[1]

- Isolated form – A sporadic disorder with no other associated conditions; this is the most common type (constituting about 66% of the cases)
- Associated/syndromic form – Accompanied with several syndromes such as hemihypertrophy of part or all of the half of the body, epidermal nevus syndrome, for example, Proteus syndrome, Klippel–Trenaunay–Weber syndrome, hypomelanosis of Ito, neurofibromatosis, tuberous sclerosis, Aicardi syndrome, and Hirschsprung's disease
- Total HME – Total HME was first described by Habervorden[7] in 1923, and it is the rarest form involving the enlargement of the ipsilateral cerebellum and brainstem. It could be isolated or associated with other syndromes.

Classification is important because investigation, management, and prognosis depend on the type of HME, which is classified into three types. The presented case had an isolated form of the abnormality, which is the most common presentation.

Clinically, epilepsy is a common manifestation occurring in more than 90% of the cases of HME.[3] Patients often present with hemiparesis, hemianopia, psychomotor retardation, focal and generalized infantile spasms, focal motor deficits, intractable seizures, developmental delays, macrocephaly, or normocephaly.[1]

Diagnosis is made via neuroimaging.[2] MRI is the imaging modality of choice for its diagnosis in infants and children due to its nonionizing nature. The diagnosis can be made *in utero* by an ultrasound scan of the cranium and also by MRI. Other imaging modalities are positron emission tomography and Computed axial tomography (CAT).[4]

In the MRI/CT findings, the main abnormalities seen are unilateral enlargement of one or part of the cerebral hemispheres with a thickened cortex.[1,4] Other features are broad gyri, polymicrogyria, or agyria; shallow sulci; indistinct gray/white differentiation; increased volume; and T2 signal of white matter. Neuronal heterotopia, ipsilateral ventriculomegaly with straightening of the frontal horn, and an "occipital sign" due to displacement of the occipital lobe across the midline occur. Gliosis and calcifications with basal ganglia and internal capsule abnormalities are also the common findings.

Management is mainly to control the seizures. Medical management for the control of seizures is often difficult with surgery, which is indicated for most cases. Hemispherectomy, which is removal of the affected hemisphere, is indicated in severe refractory cases.[8] Our patient was placed on medical management due to nonexpertise in carrying out any complex cranial surgical intervention in our environment.

Conclusion

HME is a rare congenital cortical abnormality. However, a good knowledge of its imaging features is required to identify this rare condition.

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Conflicts of interest
There are no conflicts of interest.

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