Case 14735

Eurorad

Hemimegalencephaly

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ISSN: 1563-4086
Section: Neuroradiology
Area of Interest: Neuroradiology brain
Procedure: Diagnostic procedure
Imaging Technique: CT
Imaging Technique: MR
Special Focus: Seizure disorders Case Type: Clinical Cases
Authors: Huapaya Janice, Donato Angel, Figueroa Ramon E.
Patient: 4 months, male

Clinical History:

4-month-old male patient with increasing seizure frequency.
Imaging Findings:

Brain CT: There is asymmetric enlargement/dysmorphism of the left hemisphere. The falx cerebri is shifted to the right of midline. There is irregular contour and decreased sulcation throughout the left hemisphere, suggesting diffuse pachygyria/polymicrogyria. There is asymmetric white matter hypodensity throughout the left hemisphere, which suggests abnormal myelination. The body of the left lateral ventricle is relatively enlarged.
There are multiple dystrophic calcifications throughout the left hemisphere, more confluent within the left anterior frontal subcortical white matter (Fig. 1)
Brain MRI: Significant asymmetric enlargement with dysmorphism of the left cerebral hemisphere. Diffuse loss of sulcation in association with diffuse pachygyria/polymicrogyria. There is diffusely abnormal myelination within the left hemisphere. 3D SWAN images demonstrate diffuse, irregular magnetic susceptibility artefact throughout the subcortical and deep white matter of the left hemisphere, corresponding to dystrophic calcifications on CT. (Fig. 2)
Discussion:

Hemimegalencephaly (HME) is a rare condition characterized by overgrowth of one cerebral hemisphere, with different degrees of tissue impairment. It can happen as an isolated finding or as part of a syndrome, such as hypomelanosis of Ito, tuberous sclerosis complex, epidermal nevus syndrome or Klippel–Trenaunay syndrome [1]. HME is typically associated with the triad of epilepsy, global developmental delay and contralateral motor deficit. The aetiology of HME remains unknown. It is generally assumed that HME results from abnormal neuronal and glial proliferation or apoptosis [2]. This pathology does not affect the whole hemisphere; in general, the posterior regions (parietal, occipital, and posterior aspect of the temporal lobe) are more frequently involved, compared with frontal or anterior aspect of the temporal lobes. [2]
CT findings are asymmetric unilateral enlarged cerebral hemisphere and hemicranium. Posterior falx and occipital pole "swing" to the contralateral side. Affected lateral ventricle is large with abnormally shaped frontal horn. There are also dystrophic calcifications within white matter and thickened cortex [1].

MR findings are thickened cortex, frequently increased WM signal. There is "accelerated myelination" for age and disorganized heterotopic neurons causing T1 shortening. The lateral ventricle is usually large and the frontal horn is pointed. Pachygyria, polymicrogyria is present. Margins between grey-white matter are often blurred. Diffusion

tensor images can show abnormal fibre tracts connecting hemispheres. Affected hemisphere may have bizarre enhancement [1].

Epilepsy is the most common and severe neurologic manifestation in patients with HME. The epilepsy pattern in HME can be partial seizures or may present as spasms in epileptic encephalopathy. Mental retardation and contralateral hemiparesis are associated with early epilepsy, which is highly resistant to medical management, often mandating surgery [3]. Hemispherectomy, originally described as the anatomical removal of one of the hemispheres, had been the treatment of choice in catastrophic epilepsy as in HME [4]. Surgical approach has now progressed to functional hemispherectomy or hemispherotomy, the goal and result remaining as hemispheric disconnection. Anatomic hemispherectomy, peri-insular hemispherectomy, modified lateral hemispherotomy and vertical parasagittal hemispherotomy have all been reported to be efficient in seizure control as well as in preventing further additional cognitive injury and developmental delay. [3]

Brain MRI is thus the gold standard diagnostic tool. It is very important to identify the abnormal tissue organization to make the correct diagnosis. Different degree of changes in MRI T1 and T2 signal intensity reveal the WM abnormalities and are the most important and constant sign. [2]
Differential Diagnosis List: Hemimegalencephaly, Focal cortical dysplasia, Rasmussen encephalitis, Tuberous sclerosis, Infiltrating glioma in gliomatosis pattern

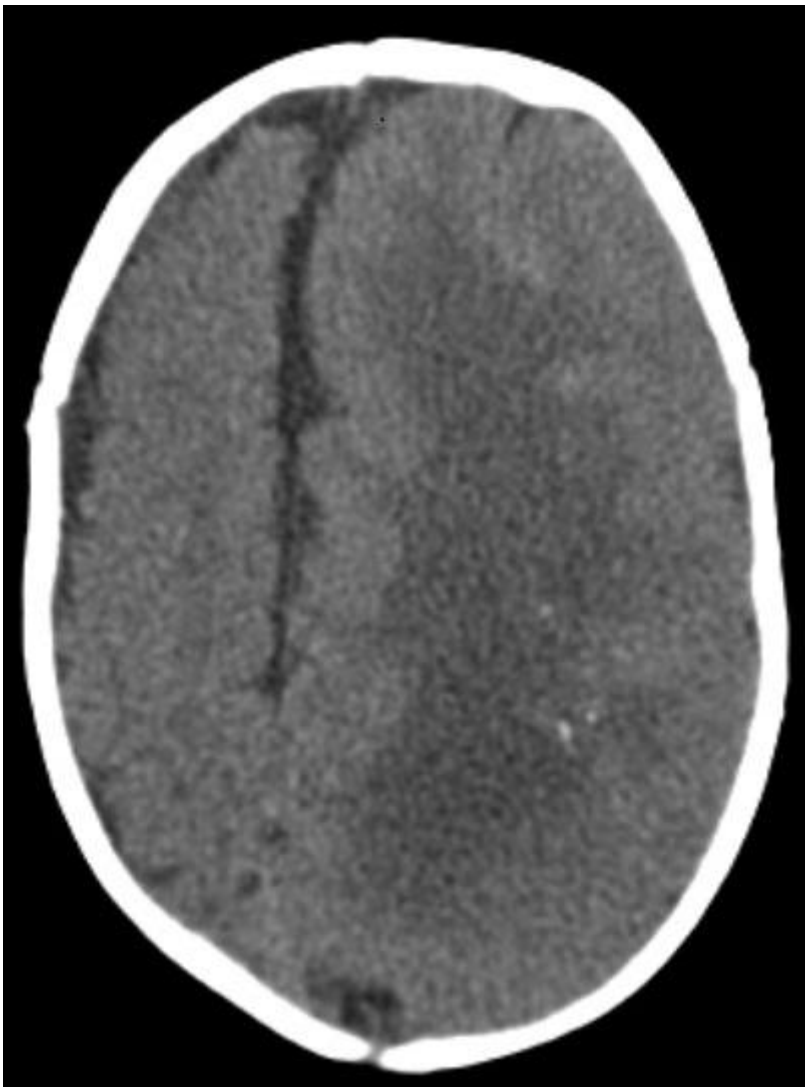
Final Diagnosis: Hemimegalencephaly

References:

Kristin M. Ikeda ?, Seyed M. Mirsattari (2017) Evolution of epilepsy in hemimegalencephaly from infancy to adulthood. elservier 7: 45–48. (PMID: 28377884)
Oikawa T, Tatewaki Y, Murata T, Kato Y2, Mugikura S, Takase K, Takahashi S (2015) Utility of diffusion tensor imaging parameters for diagnosis of hemimegalencephaly. Neuroradiology journal 28(6):628-33 (PMID:26481187)
Shilpa D Kulkarni, Chandrashekhar E Deopujari, Varsha A Patil, Rafat J Sayed (2015) Hemispherotomy in an infant with hemimegalencephaly. ediatric neurosciences Vol: 10 Issue : 2 Page: 188-192 (PMID:26167233)
VV Vaishnavathi, Rajeswaran Rangasami, Sudarshan Suresh, Indrani Suresh (2015) Fetal hemimegalencephaly. Neurology india Vol: 63 Issue: 4 Page: 636-637 (PMID: 26238918)

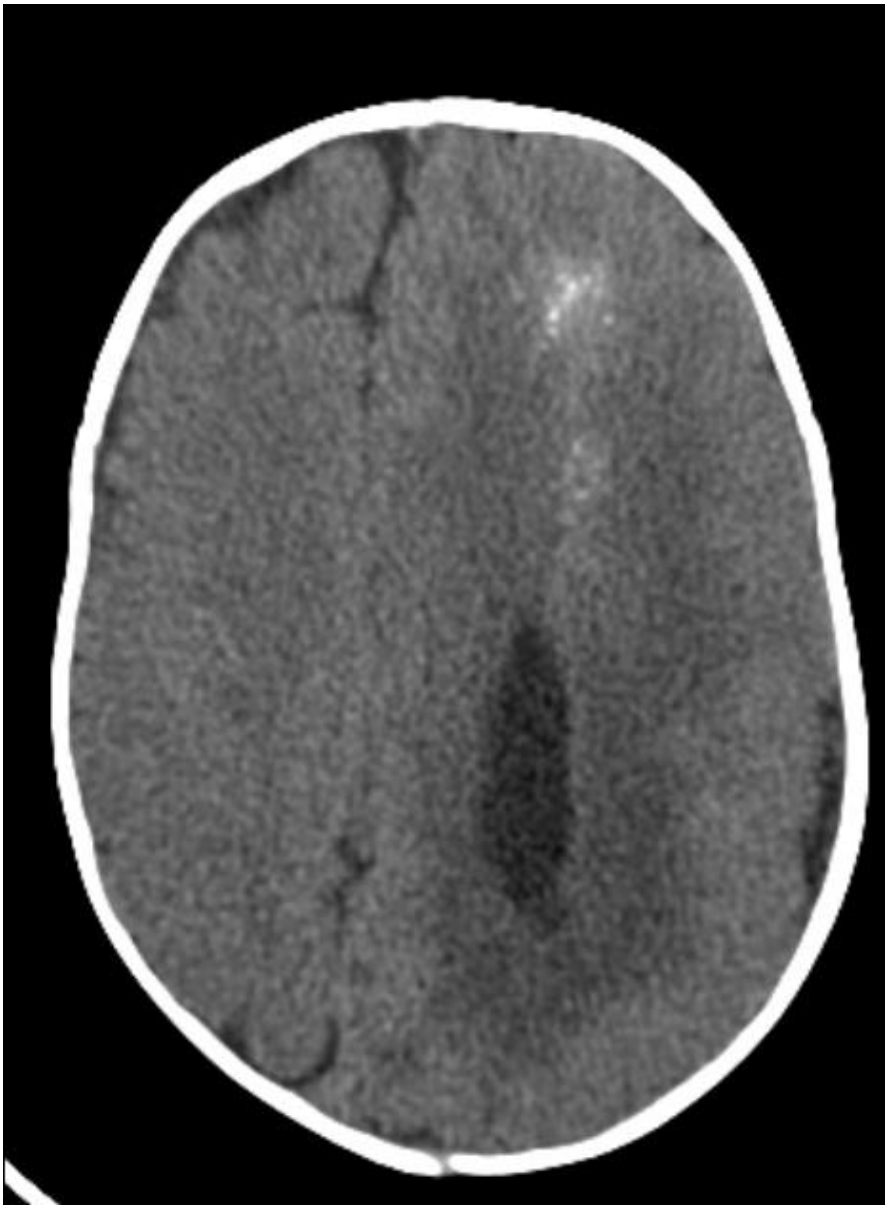
Figure 1

a



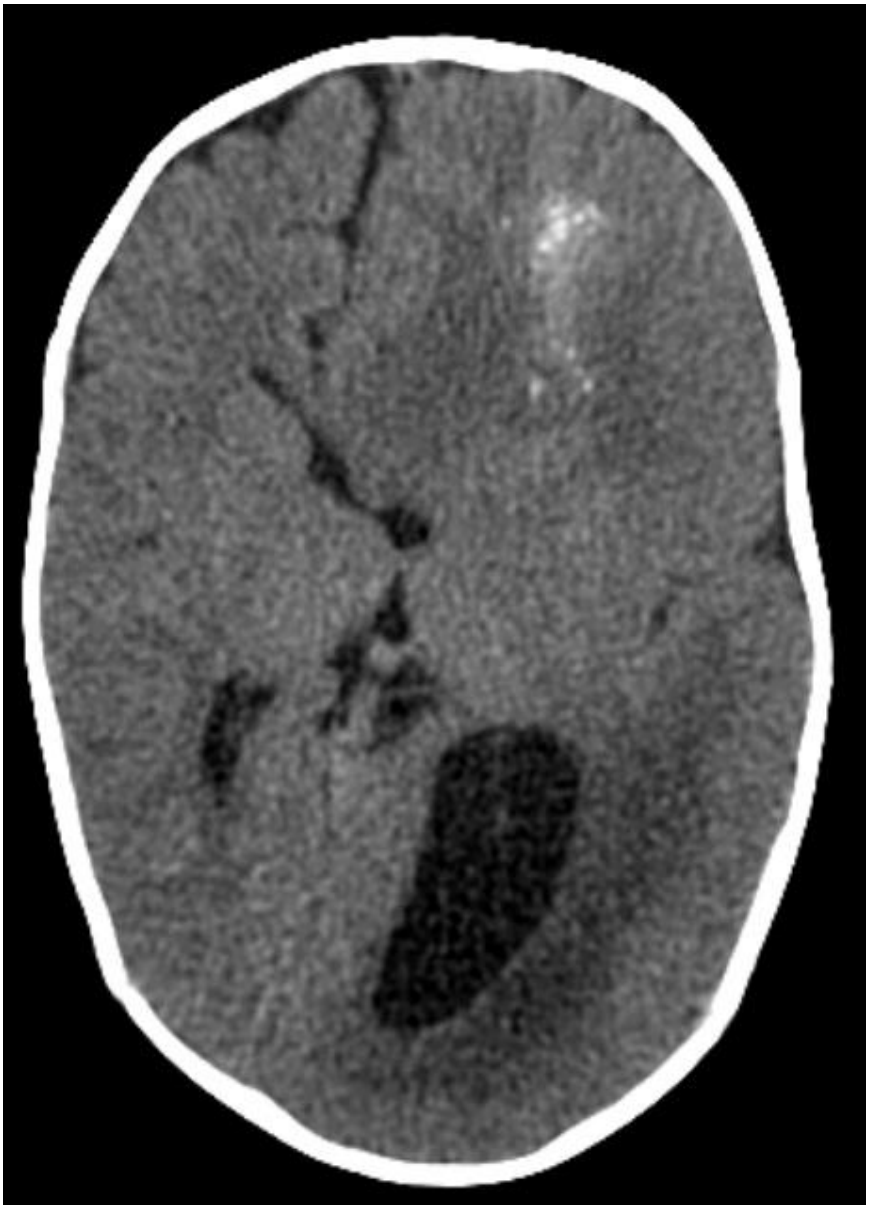
Description: Brain CT shows left hemispheric enlargement/ dysmorphism with falx cerebri shifted to the right. There is irregular contour and decreased sulcation throughout the affected hemisphere. Notice dystrophic calcifications in supraventricular white matter. **Origin:** augusta university

b



Description: Multiple dystrophic calcifications throughout the left hemisphere, more confluent within the left anterior frontal subcortical white matter, but extending to the supraventricular parietal centrum semiovale. **Origin:** augusta university

c



Description: Multiple dystrophic calcifications throughout the left hemisphere, more confluent within the left anterior frontal subcortical white matter, but extending to the supraventricular parietal centrum semiovale. **Origin:** augusta university

Figure 2

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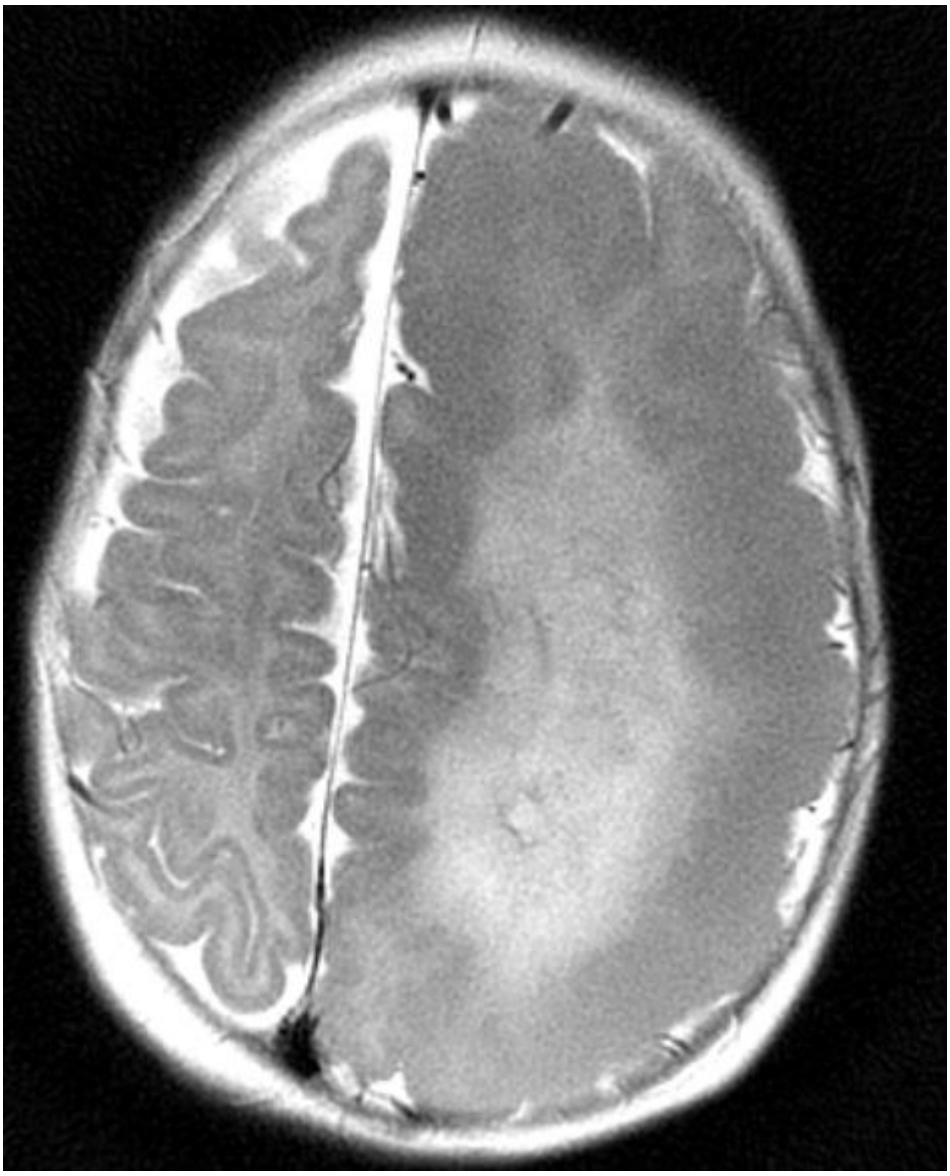
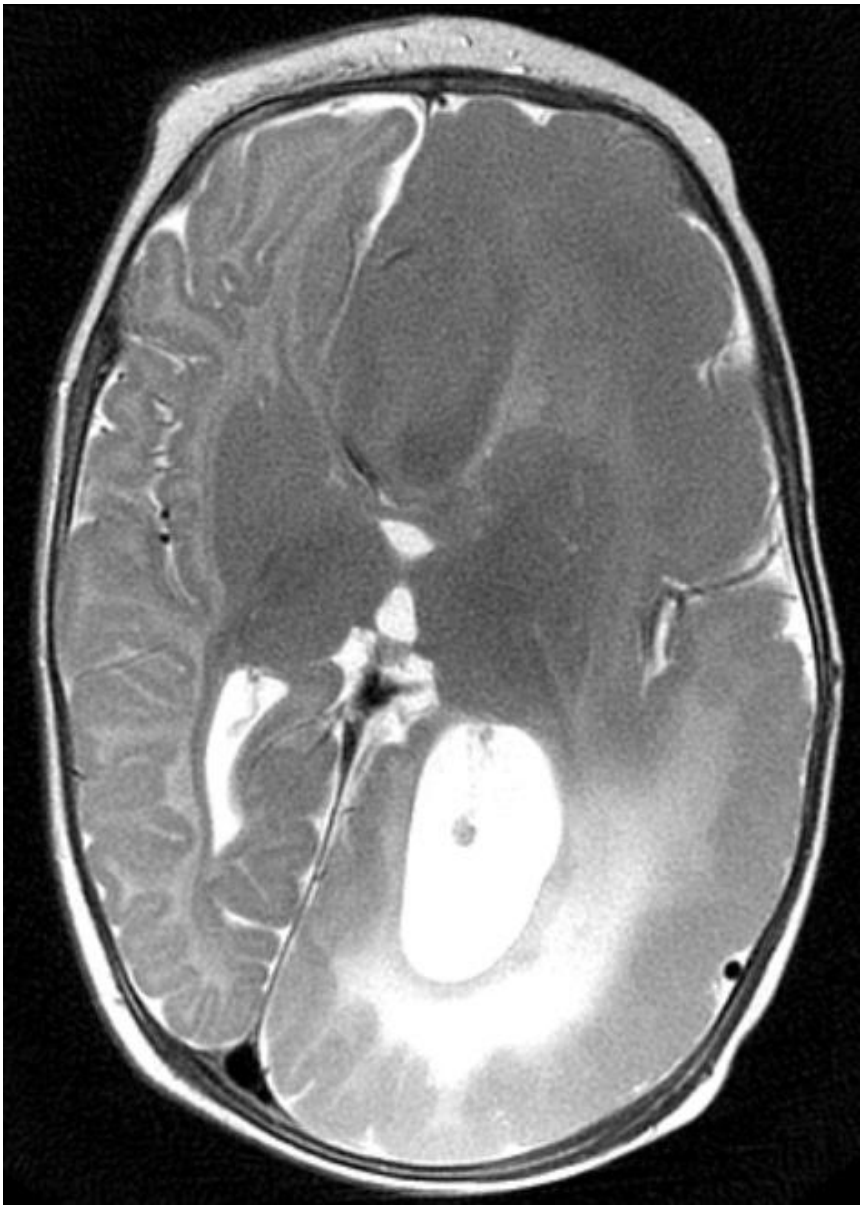


Description: MRI brain shows significant dysmorphic enlargement of the left cerebral hemisphere. Diffuse pachygyria/ polymicrogyria results in loss of sulcation, with abnormal left hemispheric myelination. **Origin:** augusta university

b



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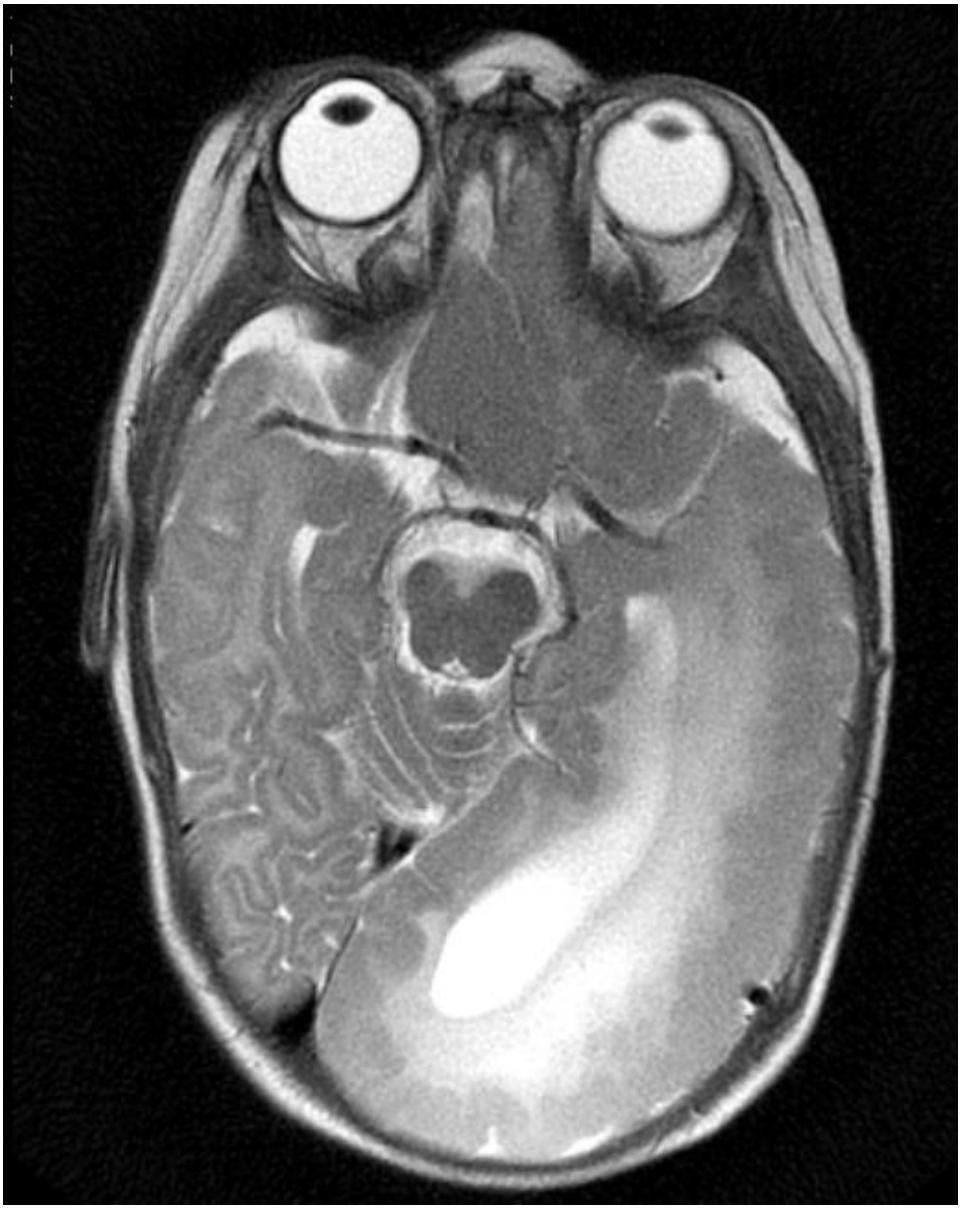
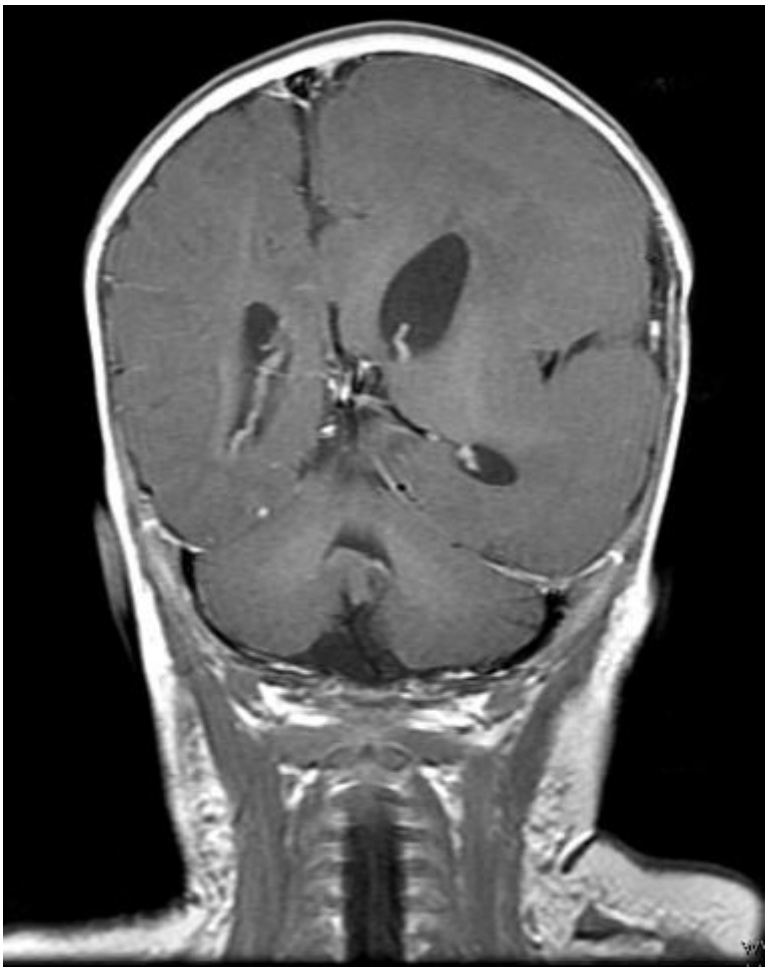


Description: MRI brain shows significant dysmorphic enlargement of the left cerebral hemisphere. Diffuse pachygyria/ polymicrogyria results in loss of sulcation, with abnormal left hemispheric myelination. **Origin:** augusta university

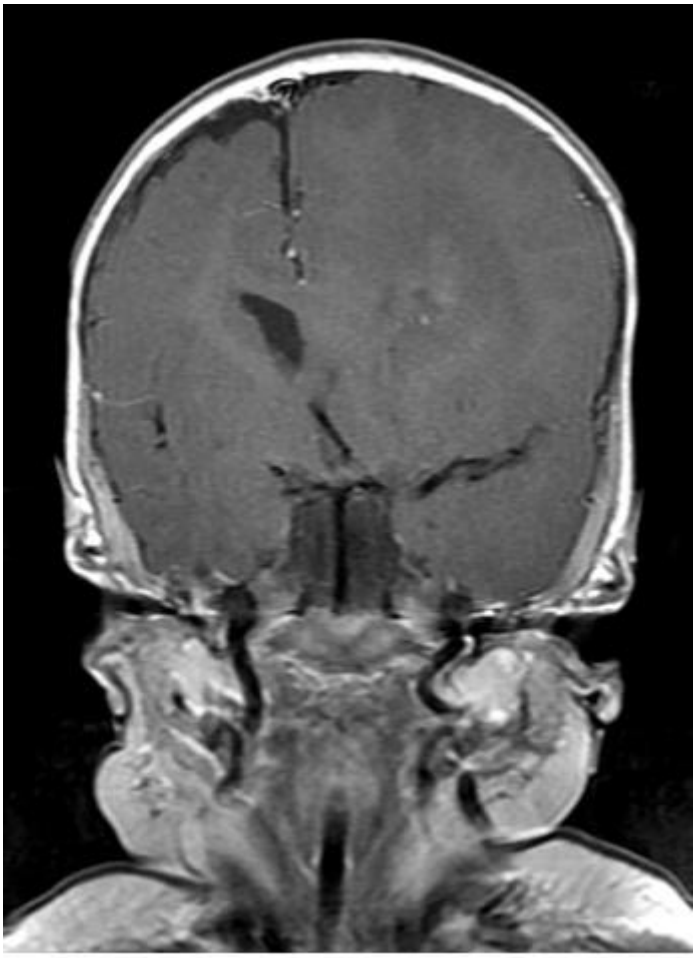
Figure 3

a



Description: MRI T1 coronal brain shows to better extent the volume discrepancy between hemispheres with dysmorphic left hemisphere features with loss of sulcation. **Origin:** augusta university

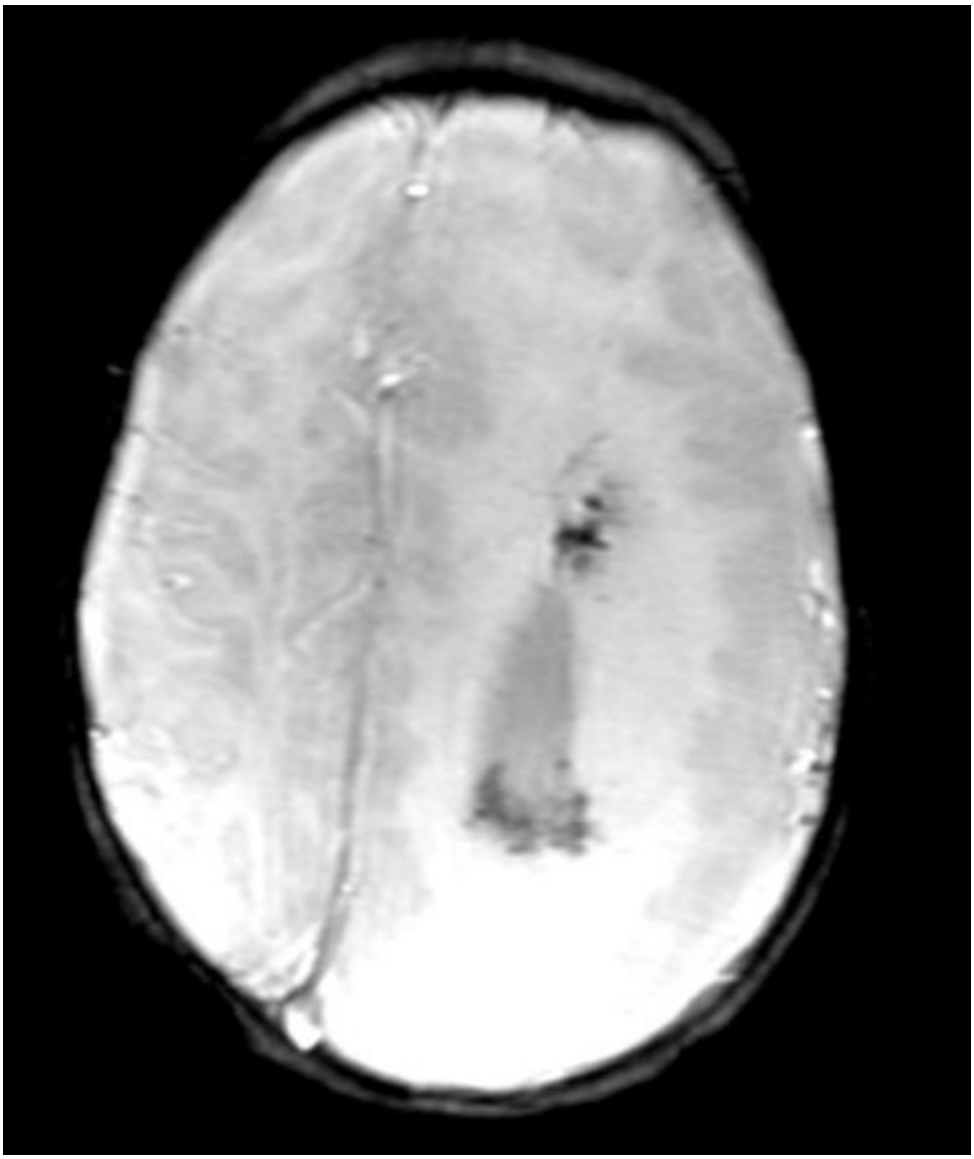
b



Description: MRI T1 coronal brain shows to better extent the volume discrepancy between hemispheres with dysmorphic left hemisphere features with loss of sulcation. **Origin:** augusta university

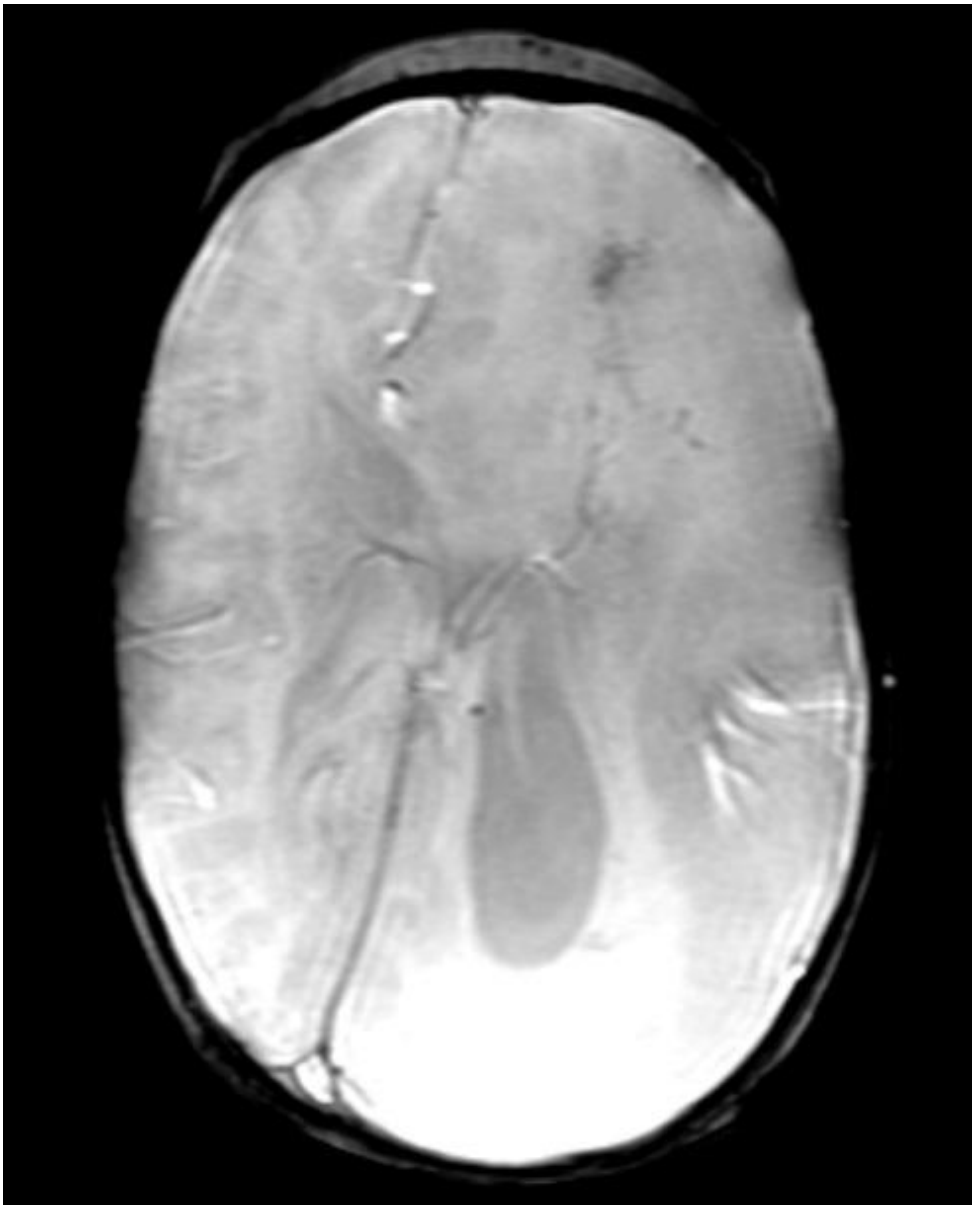
Figure 4

a



Description: 3D susceptibility (SWAN) images demonstrate diffuse, irregular magnetic susceptibility artifact throughout the subcortical and deep white matter of the left hemisphere, corresponding to dystrophic calcifications seen on CT. **Origin:** augusta university

b



Description: 3D susceptibility (SWAN) images demonstrate diffuse, irregular magnetic susceptibility artifact throughout the subcortical and deep white matter of the left hemisphere, corresponding to dystrophic calcifications seen on CT. **Origin:** Augusta university