"Batwing\' sylvian fissure

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Section: Neuroradiology
Case Type: Anatomy and Functional Imaging

Patient: 1 years, male

Clinical History:

A 1 year old boy presented with history of seizures and delayed attainment of developmental milestones. He was born to non-consanguineous parents and had no history of any perinatal complications. Physical examination revealed macrocephaly, while the routine laboratory investigations were within normal limits. Biochemical investigations subsequent to neuro-imaging clinched the diagnosis.

Imaging Findings:

CT scan of the brain demonstrated fronto-temporal atrophy with widening of adjacent CSF spaces and generalized prominence of the ventricular system (Fig 1). The distinct imaging finding noted was the presence of widely open opercula (sylvian fissures) in the form of ‘batwings’ (Fig 2,3). Axial T1- and T2-weighted MRI demonstrated subdural bleed along the left fronto-parietal cerebral convexity (Fig 4,5). Diffuse abnormal high-signal intensity was seen in the supratentorial white matter on T2-weighted images along with few focal T2-weighted signal alterations within the basal ganglia (Fig 5). Based upon the characteristic neuroimaging findings and relevant clinical history, diagnostic possibility of glutaric aciduria type-I was considered which was subsequently confirmed on biochemical analysis.

Discussion:

Glutaric aciduria type-I is a rare autosomal recessive disorder caused by the deficiency of a mitochondrial enzyme glutaryl-CoA dehydrogenase. This enzyme deficiency is responsible for the improper breakdown of the aminoacids resulting in an elevated plasma and urine level of glutaric acid.

Glutaric acid accumulation in the body is responsible for neurotoxicity in the basal ganglia and fronto-temporal cortex. Pathologically, there is extensive neuronal loss caused in the basal ganglia and fronto-temporal region with severe spongiform changes in the white matter. This neurodegeneration may manifest as progressive dystonia, hypotonia, permanently impaired speech or seizures in an infant or toddler. A prominent clinical feature of these patients is the presence of macrocephaly. Clinical features are typically preceded by an acute encephalopathic illness; hence many a times these cases are mistaken for acute encephalitis, Reye syndrome or vaccine induced encephalopathy.

Neuroimaging serves as a useful tool, many a times providing the first clue to the diagnosis. The earliest imaging finding is frontotemporal cerebral atrophy. The most striking finding on brain imaging is the presence of very wide CSF spaces anterior to the temporal lobes and along the sylvian fissures. The appearance of widely dilated opercula has been likened to ‘wings of a bat’. This ‘Batwings dilatation’ of sylvian fissure was first described as a characteristic imaging finding in glutaric aciduria in 1987. Although the finding of wide opercula suggests glutaric aciduria type I, the finding is by no means specific, as this imaging appearance may also be encountered in severe
developmental delay and idiopathic external hydrocephalus. But if this 'Batwings dilatation' of sylvian fissure is associated with concomitant basal ganglia lesions, it is considered almost pathognomonic of glutaric aciduria type I. Basal ganglia changes include bilateral basal ganglia atrophy and T2-hyperintensity. White matter changes may be seen as diffuse low density on CT or diffuse T2-weighted hyperintensity on MRI. Rupture of bridging veins may lead to subdural bleed especially along the frontotemporal cerebral convexity, which may lead to the suspicion of non-accidental injury.

To conclude, the finding of 'Batwings dialatation' of sylvian fissures and wide CSF spaces anterior to temporal lobes in a child should alert the radiologist of the possibility of Glutaric aciduria type I especially if its associated with other ancillary imaging findings such as basal ganglia lesions, subdural bleed or diffuse white matter changes.

**Differential Diagnosis List:** Glutaric aciduria type-1, Bilateral temporal fossa arachnoid cysts (on imaging), Severe developmental delay, Idiopathic external hydrocephalus

**Final Diagnosis:** Glutaric aciduria type-1

**References:**


**Description:** Axial CT scan shows bi-frontal cerebral atrophy with associated subarachnoid CSF space and ventricular prominence. **Origin:**
Description: Widely open opercula are evident on both sides (right>left). Origin:
Figure 3

Description: Widely open sylvian fissures have been likened to batwings. This has been considered the most striking finding on neuroimaging in glutaric aciduria type I. Origin:
Description: Subdural hygroma can be seen along the left fronto-parietal cerebral convexity. Origin:
**Description:** Diffuse white matter hyperintensity & focal signal alterations within the basal ganglia can be seen on this T2-w MRI. Also seen are left fronto-parietal subdural bleed, bi-fronto-temporal atrophy, enlarged opercula and subtle basal ganglia atrophy. **Origin:**