

Case Report

Corpus Callosal Lipoma with Agenesis of Corpus Callosum- A Case Report

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Abstract:

More than one half of intracranial lipomas are associated with varying degrees of brain malformations. Associated anomalies include agenesis of surrounding tissues, frontal bone defects or facial dysplasia and cerebral vascular defects. Agenesis of corpus callosum can be associated with a midline lipoma with or without calcification of its periphery. This developmental abnormality has an embryological basis. We observed this in 1 case, and as this is uncommon, are reporting this.

Key words: Corpus Callosum, Lipoma, Agenesis.

Introduction:

Agenesis of corpus callosum (ACC) is a rare birth defect, in which there is complete or partial absence of corpus callosum. It occurs when the corpus callosum, the band of white matter connecting the hemispheres in the brain, fails to develop normally, typically during first trimester of pregnancy. Intracranial lipomas represent rare congenital malformations, accounting for less than 1% of intracranial tumours. They are thought to originate from abnormal differentiation of the meninx primitive, a mesenchymal derivative of the neural crest¹. Intracranial lipomas may present with symptoms such as headache, seizures, local mass effect or may be diagnosed incidentally during evaluation following trauma².

Causes of ACC

In most cases the cause of ACC is unknown. However agenesis of corpus callosum can be inherited as autosomal recessive trait or an X-linked dominant trait. This disorder may also be due in part to an infection or

injuries during pregnancy (Intrauterine), prenatal toxic exposures, structural blockage by cyst or lipoma and other metabolic disorders.

Signs and symptoms of ACC

Signs and symptoms of ACC and other callosal disorder vary greatly among individuals. However some characteristics common in individuals with callosal disorders include vision impairments, low muscle tone, poor motor coordination, delays in motor milestones such as sitting and walking, low perception of pain, delayed toilet training, chewing and swallowing difficulties, difficulty in complex problem solving (Cognitive disability), social difficulties (missing subtle social cues) even when their IQ is normal. Other characteristics sometimes associated with ACC include seizures, spasticity, early feeding difficulties, hearing impairments, abnormal head or facial features and a mental handicap.

Investigations:

Callosal disorders can be diagnosed through an MRI, CT scan, Sonography, Prenatal Ultrasound or prenatal MRI.

Treatment:

There is currently no specific medical treatments for ACC, although seizures if present need medical management. In the rare cases where a cranium bifidum is present then surgical repair may be necessary. Individual with ACC may benefit from a

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range of developmental therapies and educational support. It is important to consult with a variety of medical, health educational and social work professionals.

Prognosis

Prognosis is good; but the degree of disability is variable, ranging from severe neurological dysfunction to average intelligence and normal lives³.

Case report:

A 25 years old female was involved in a road traffic accident. She was brought to emergency department after a high speed frontal impact against road. She was treated according to advanced trauma life support guideline. She also had history of occasional seizure for last 3 years for which she did not receive any treatment. Owing to the fact that the patient had previous seizure that could not be explained and trauma in road traffic accident -a brain CT was ordered which shows -acute intracerebral hematoma in left fronto-parietal lobe and a lipoma along septum pellucidum and mild extra calvarial soft tissue swelling. On the basis of CT report MRI of brain was advised. MRI shows -Corpus callosum agenesis with tubulonodular variety of corpus callosal lipomas and subacute hemorrhage of left frontal lobe.

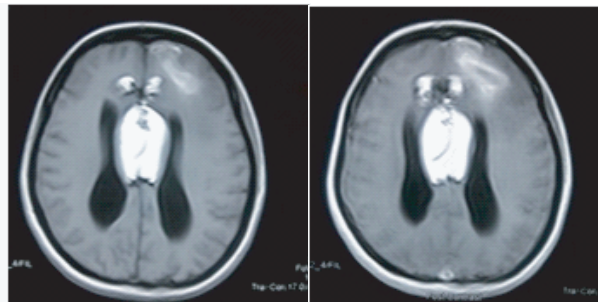


Figure 1: T1WI axial MR image show parallel lateral ventricle due to corpus callosal agenesis giving the racing car sign appearance and increase signal intensity corpus callosal lipoma. Post contrast T1WI MR image show no enhancement of lipoma. There is a high signal intensity area in left frontal lobe due to haemorrhage.

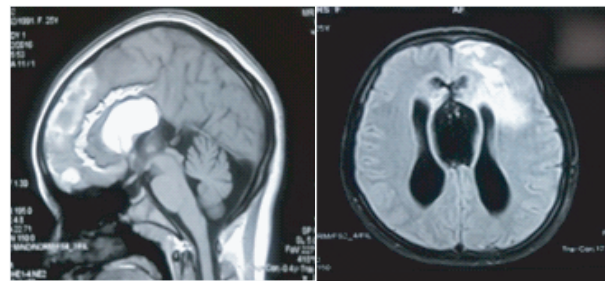


Figure 2: T2WI Sagittal. FLAIR MR demonstrates hypoplasia of genu, body, splenium & rostrum of corpus callosum (corpus callosal dysgenesis) with a tubulonodular variety of corpus callosal lipoma.

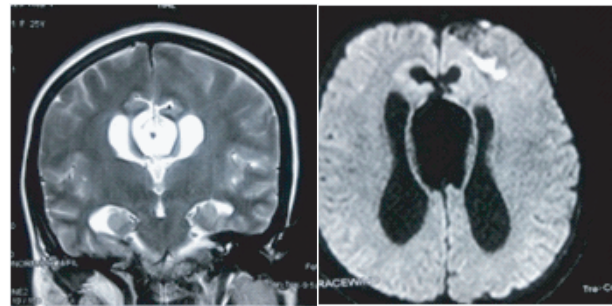


Figure 3: This case demonstrates the moose head sign (also known as viking helmet sign) with tubulonodular variety of corpus callosal lipoma in coronal T2WI MR image and lipoma is suppressed in STIR MR image.

Discussion:

Agenesis of the corpus callosum may be complete or partial and represents an in utero developmental anomaly. It may be as uncommon as 1:20,000 according to autopsy series.

The development of the corpus callosum occurs between the 12th and 16-20th weeks of gestation. It begins with the genu and then continues posteriorly along the body to the splenium. The rostrum is the last part to be formed.

Myelination of the corpus callosum occurs in the opposite direction, from the splenium forwards.

Agenesis is a result of an insult occurring at approximately 8-12 weeks gestation resulting in failure to form the corpus callosum. The white matter tracts which usually cross the midline instead are oriented vertically, separating the lateral ventricles widely, in a racing car sign configuration. These bundles of white matter are known as Probst bundles. The anterior commissure is usually present and often enlarged. The hippocampal formations are usually hypoplastic, with resultant dilatation of the temporal horns of the lateral ventricles.

Agenesis (which may be complete or partial) is a result of encephalomalacia secondary to toxic, ischaemic or traumatic events. Isolated partial agenesis of the corpus callosum is often asymptomatic. The clinical picture in other cases is dictated by the associated abnormalities that are frequently found, especially in agenesis.

Corpus callosum lipomas are fat-containing lesions occurring in the interhemispheric fissure closely related to the corpus callosum, which is often abnormal. It is the most common location for an intracranial lipoma. On imaging, they can be identified in two different

morphologies: tubulonodular, which is the most common, and usually presents as a rounded anterior nodular mass; and curvilinear, which is usually thin, elongated and curvilinear along the corpus callosal margin. They show fat density on CT (-80 to -110HU) and a fat signal on MRI, which is characterised by a markedly elevated T1 signal and high T2 signal, without contrast enhancement^{4,5}.

Intracranial lipomas are thought to be benign, slow-growing, congenital hamartomatous conditions, which are very rare⁴. Approximately 50% of patients of corpus callosal lipomas present with seizures. The tubulonodular variety is usually associated with more severe and extensive abnormalities and thus is more frequently symptomatic⁶.

The pathogenesis of a corpus callosum lipoma is considered to be the result of an abnormal persistence and differentiation of the meninx primitiva into lipomatous tissue. Typically resorption occurs between the 8th and the 10th week of gestation^{4,7}.

Tubulonodular corpus callosum lipomas are the more common variety. They are rounded or lobular and usually measure >2 cm in thickness. They are anteriorly situated and are associated with extensive callosal and often fronto-facial anomalies. The tubulonodular variety can extend into the choroid plexus lateral ventricles. Curvilinear corpus callosum lipomas are usually thin, elongated and curvilinear along the corpus callosum margin. They usually measure <1 cm in thickness and are more posteriorly situated. The corpus callosum is only mildly hypoplastic.

Our patient had classical features of agenesis of corpus callosum, viz, high riding third ventricle, separated and parallel lateral ventricles, colpocephaly and spoke wheel appearance of sulci over medial surface of cerebral hemispheres.

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