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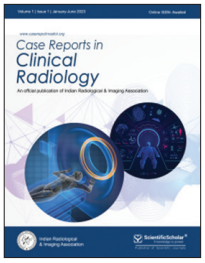

Case Report

A unique case of agenesis of corpus callosum with concomitant midline teratoma and dorsal interhemispheric cyst

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ABSTRACT

Midline interhemispheric cyst with corpus callosal agenesis is a well recognized condition and has been associated with various genetic conditions. Associated extraaxial tumors are uncommon with no proven genetic association. We describe a rare case of corpus callosal agenesis with a dorsal interhemispheric cyst and an extraaxial teratoma in the basifrontal region and review its imaging features along with the etiopathogenesis of concurrence of such condition.

Keywords: Corpus callosal agenesis, Interhemispheric cyst, Midline teratoma

INTRODUCTION

The corpus callosum is the largest interhemispheric commissure of the human brain.[1] Corpus callosum dysgenesis includes agenesis and hypogenesis and association with an interhemispheric (IH) cyst is a rare entity. Antenatal diagnosis of different clinical syndromes associated with agenesis of corpus callosum (CCA) such as Chiari or Dandy–Walker malformation is routinely done; however, extra-axial brain tumors are an extremely rare association with a relative dearth of reported cases. Radiological imaging shows a fat-containing extra-axial lesion in basifrontal region with internal calcifications and enhancing soft tissues. We describe a rare case of corpus callosum agenesis with IH cyst as well as an extra-axial teratoma in the basifrontal region along with a brief review of its imaging features and pathogenesis.

CASE REPORT

A 3.5-year-old male child presented in the Pediatric Outpatient Department of a tertiary hospital with gross motor developmental delay and inability to walk. Birth history was insignificant. No antenatal ultrasounds were done. Physical examination revealed macrocephaly, hypertelorism and high arched palate. Vision and hearing were unremarkable.

CE-magnetic resonance imaging (MRI) of the brain was done which revealed a multiloculated midline extra-axial solid-cystic lesion epicentered in the basifrontal region of the anterior cranial fossa with mixed MR signal characteristics and multifocal enhancing soft tissue. Axial T1W

[Figure 1a], T2W [Figure 1b], Post contrast [Figure 1c] and susceptibility weighted image [Figure 1d] shows T1 and T2 WI hyperintense areas showing susceptibility artifacts representing both fat and calcification within the lesion, as was correlated on non-contrast computed tomography head [Figure 2a]. There was complete CCA with widely separated bilateral lateral ventricles [Figure 1e]. A large cerebrospinal fluid (CSF) intensity dorsal IH cyst was seen in the right frontoparietal paramedian location without any communication to the ventricular system [Figure 1e]. An arachnoid cyst was seen in the left temporal fossa [Figure 1f]. Furthermore, bilateral partial hippocampal inversion, platybasia, absent bilateral nasal bones, and high arched palate were seen with thinning of the right palatine bone. Intraoperative findings revealed a well-defined solid-cystic moderately vascular basifrontal mass seen adhered to A3, A4 segments of the right anterior cerebral artery, and right optic nerve [Figure 2b]. Histopathology showed elements of ectoderm, mesoderm, and endoderm without any atpia with a final diagnosis of mature cystic teratoma. The patient was discharged without any significant post-operative complications.

DISCUSSION

Corpus callosum is the largest interhemispheric commissure of the human brain[1] playing a pivotal role in signal transmission across the two cerebral hemispheres.[2] It has four segments: The rostrum, genu, body, and splenium with an intervening isthmus. Corpus callosum develops from the sulcus medianus telencephalii medii formed in the depression of focal thickening in lamina terminalis. Massa commisuralis shaped along the floor of IH fissure forms the bed for ingrowing crossing axonal fibers from either cerebral hemisphere, later forming the corpus callosum. The mature corpus callosum is developed by the 20th week of gestation.[3] As myelination proceeds, both length and width of corpus callosum increases with visible thickening of the genu and splenium. By 10 months of age, the overall appearance resembles that of a normal adult. Dysgenesis of corpus callosum is a disturbance of axonal conditioning and path finding in the commissural development consequential to insult at the stage of massa commisuralis formation or development of sulcus medianus telencephalii medii

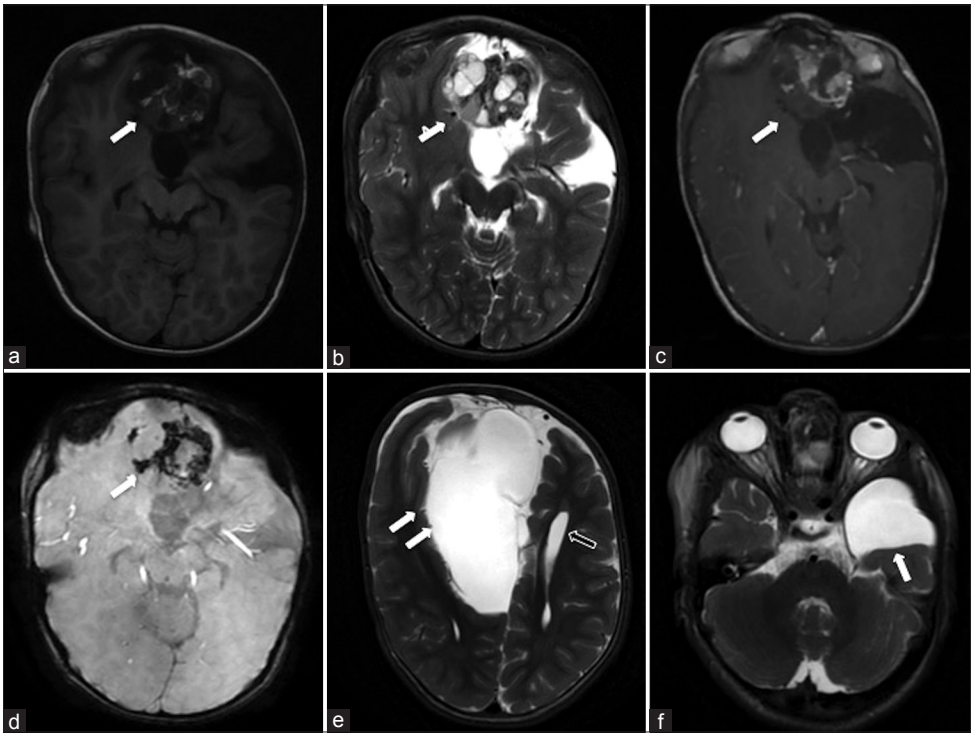

Figure 1: Axial T1W (a), T2W (b), T1WI post-contrast (c), and Susceptibility weighted images (d) show well-defined solid cystic lesion with T1 hyperintense and T2 hypointense components with hypointensity of SWI images. Enhancing soft-tissue signal intensity components also seen within. Axial T2WI (e) images of brain show parallel orientation of with midline sagittal sections suggestive of corpus callosal agenesis. A dorsal extra-axial CSF signal intensity cyst (solid white arrows in e) is seen along the falx cerebri communicating with third ventricle. Caudal axial T2WI sections (f) show a CSF signal intensity extra-axial arachnoid cyst (white arrow) in the left anterior temporal fossa causing mass effect over underlying anterior temporal lobe. T1W1: T1-weighted imaging, T2W2: T2-weighted imaging, SWI: Susceptibility-weighted imaging, and CSF: Cerebrospinal fluid.

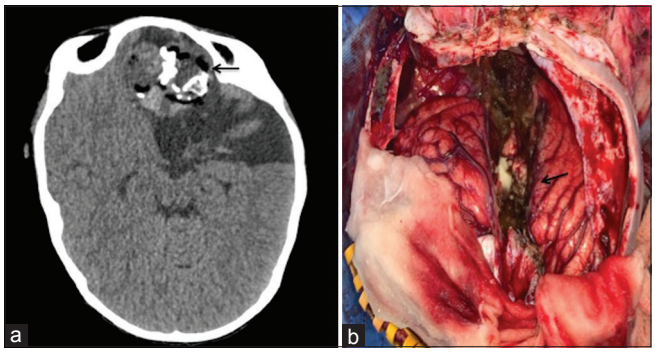

Figure 2: Axial NCCT Brain (2a) section corresponding with the MRI sections reveals presence of fat as well as calcifications in the areas of hypointensity on SWI [Figure 1d]. Intraoperative image [Figure 2b] shows presence of cheesy material and soft tissue mass in the midline. NCCT- non-contrast computed tomography and SWI-Susceptibility weighted imaging.

and is neither fatal nor results in substantial neurological deficits in all cases. As CCA is an aberration in ontogenesis, it is frequently associated with various other cerebral malformations with multitude of known syndromic associations and genetic mutations.[4]

In CCA, there is anomalous longitudinal orientation of the callosal bundle fibers along the walls of lateral ventricles with every so often presence of mesodermal tissue within the IH fissure. This abnormally located mesodermal tissue may lead to conception of various midline tumors such as lipoma (being the most commonly associated lesion), teratoma, fibrous tumor, or meningioma. The occurrence of midline lipoma is common finding with presence of calcification characteristically seen in corpus callosal lipomas.[5] T1-weighted imaging (T1WI) hyperintense intralesional fat components may occasionally show susceptibility artifacts; microscopic calcifications and mineralization due to foci of osseous metaplasia, being postulated as the cause for the same.[6,7] Fat-suppressed T1WI and computed tomography scan are of utmost value in such cases to characterize the lesion.[8]

Midline IH cyst is a common association with CCA. Barkovich A J *et al*. divided CCA with IH cyst into two major groups[9] – type I, in which the IH cyst is a diverticulum of the ventricular system with communication and type 2, in which multiple cysts are present with no communication with ventricular system. Some authors believe the origin of these intracranial cysts to be controversial – that is, arachnoid, neuroepithelial, or ependymal in origin.[10] Type II D IH cyst found in our case is considerably rare and it demonstrates no communication with ventricular system and shows CSF iso signal intensity.

Concurrence of an extra-axial teratoma with IH cyst with CCA has seldom been reported in the literature. This concomitant occurrence of IH cyst as well as teratoma in CCA may be explained by the fact that both teratoma and IH cyst are developmental anomalies occurring due to abnormal differentiation of mesodermal tissue in the IH fissure. In addition, there is another arachnoid cyst in the left temporal region. Arachnoid cysts are thought to develop secondary to splitting or duplication of normal arachnoid membrane during development[11] and are symptomatic complains largely due to their mass effect on underlying brain pernchyma.

CCA is also almost always accompanied by the absence of the hippocampal commissure and hippocampal malrotation. Association with numerous genetic conditions and syndromes such as Chiari 2 malformation, Dandy–Walker spectrum, frontonasal dysplasia, median cleft face syndromes, and syndromic craniosynostoses has been described.[3]

Richeiri-Costa and Guion-Almeida reported ten sporadic cases of patients with facial midline defects, callosal agenesis, basal encephalocele, and ocular anomalies as frontonasal dysplasia spectrum.[12] Facial anomalies were variable from mild hypertelorism to median cleft lip and ocular anomalies included findings such as ptosis, orbital asymmetry, and optic disk anomalies. In our case, the patient did present with facial dysmorphic features in form of hypertelorism, absent nasal bones, and high arched palate. However, no cleft lip or palate and basal encephalocele were found. There were no cortical malformations or subcortical and subependymal heterotopias. Long and small bones of the extremities were normal.

Differential diagnosis
CCA with lipoma
Frontonasal dysplasia
Median cleft face syndromes
Acrocallosal syndromes

CONCLUSION

Co-occurrence of CCA with midline teratoma and IH cyst is a rare entity and can be attributed to embryological insult at the stage of massa commisuralis formation or development of sulcus medianus telencephalii medii and presence of aberrant mesodermal soft tissue in IH fissure. MRI plays an important role in detection of associated cerebral anomalies such as migration and sulcation abnormalities and presurgical evaluation of extents of the cystic lesions. Further, genomic work up is recommended in such cases to rule out genetic associations.

TEACHING POINTS

- Corpus callosal dysgenesis is commonly associated with other midline and hemispheric anomalies and should be keenly looked for in each case
- Presence of enhancing soft tissue within the midline lesion points toward the diagnosis of teratoma rather than a callosal lipoma as calcification can be seen in either of the lesions

- Susceptibility artifacts within a tumor should also raise the possibility of fat components within the lesion besides the usual hemorrhage and calcification.

MCQs

1. Correct sequence of embryological corpus callosal development is-
   a. Callosal commissural plate-genu-rostrum-splenium
   b. Genu-callosal commissural plate-splenium-rostrum
   c. Callosal commissural plate-rostrum-genu-splenium
   d. Callosal commissural plate-splenium-rostrum-genu

Answer Key: c

2. Various syndromes associated with CCA are all except-
   a. Frontonasal dysplasia
   b. Aicardi syndrome
   c. Wallenberg syndrome
   d. Dandy–Walker malformations

Answer Key: c

3. Calcification can be seen in –
   a. Teratoma
   b. Callosal lipoma
   c. None of the above
   d. All of the above

Answer Key: d

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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