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Case Report

Agenesis of the Corpus Callosum Associated with Persistent Primitive Olfactory Artery - Case Report and Literature Review

Abstract

Agenesis of the corpus callosum (ACC) is a congenital malformation that can occur in isolation or in association with other neurological conditions. However, to our knowledge, no cases of ACC associated with persistent primitive olfactory artery (POA) have been reported. Here we report a case involving a patient with ACC associated with persistent POA. A 57-year-old man was admitted to our hospital because of trauma to his face. Neurological findings were normal. Face computed tomography (CT) showed a mandible fracture. Brain CT and magnetic resonance imaging (MRI) demonstrated total ACC, intact anterior commissure, and no development of hippocampal commissure. CT angiography of the brain demonstrated a right persistent POA. The findings in this case suggest that CT angiography or MRI may detect persistent POA more effectively in cases involving ACC.

Introduction

Agenesis of the corpus callosum (ACC) is one of the most common human brain malformations. ACC results from commissural defects, and although it can occur for a number of reasons, including genetic, metabolic, or vascular abnormalities, in most cases, the cause remains unknown [1]. Brain anomalies and other malformations associated with ACC include lipoma of the corpus callosum, cerebellar hypoplasia, Dandy-Walker syndrome, hydrocephalus due to obstruction of the foramen of Monro or aqueduct stenosis, interhemispheric cyst, porencephalic cyst, and neuronal migration disorders [2]. Associated craniofacial dysmorphic features include hypertelorism, encephaloceles, and craniosynostosis. Commonly seen ocular anomalies include microphthalmia, coloboma, retinal lacunae (Aicardi syndrome), and optic nerve hypoplasia [2]. Cerebral vascular anomalies associated with ACC include sinus pericranii [3], meandering of the anterior cerebral artery (ACA), coursing of the ACA directly upwards in the interhemispheric fissure, and persistent falcine sinus [4].

Persistent primitive olfactory artery (POA) is a rare anomaly of the ACA frequently associated with anomia and a saccular aneurysm within a hairpin turn [5,6]. However, to our knowledge, no cases of persistent POA associated with ACC have been reported. Here we report a case of persistent POA associated with ACC and review this anomaly with a focus on the mechanism of its development.

Case Report

A 57-year-old man presented with face trauma after a bicycle accident. He complained of chin pain and headache, and had three broken teeth. Physical examination demonstrated a chin laceration and multiple wounds to the face. Neurological examination showed no abnormalities. Face computed tomography (CT) showed mandible fracture. Brain CT in the emergency department

demonstrated suspicious ACC and dilation of the atrium and occipital horns of both lateral ventricles (colpocephaly) (Figure 1). Brain magnetic resonance imaging (MRI) showed radially arrayed gyri pointing to the third ventricle, an absent cingulate gyrus, Probst bundle, total ACC, and anterior commissure (Figure 2). Brain MRI also demonstrated a suspicious persistent POA (Figure 3). Brain CT angiography showed a right persistent POA (Figure 4). Sagittal CT angiography demonstrated a highly located internal cerebral vein (Figure 5). Open reduction and internal fixation for the mandible fracture were performed on the eighth day after trauma, and the patient was discharged on the 16th day after trauma.

Discussion

Agenesis of the corpus callosum

In humans, the corpus callosum starts to develop at the estimated gestational age (EGA) of approximately 8 weeks, and it



Figure 1: Brain axial computed tomography showing suspicious agenesis of the corpus callosum (ACC) (arrow) and dilation of the atrium and occipital horns of both lateral ventricles (colpocephaly, arrowhead).

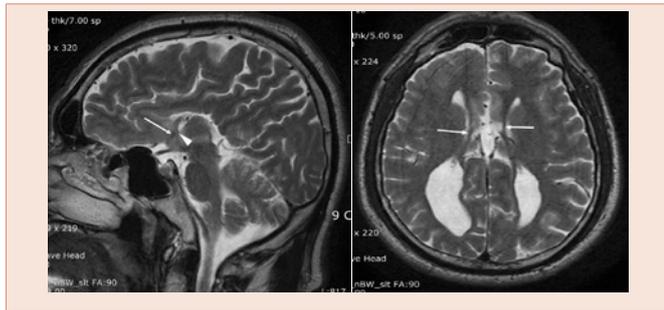


Figure 2: Brain magnetic resonance image (MRI) demonstrating radially arrayed gyri pointing to the third ventricle, definite anterior commissure (arrow), fornix (arrowhead), total ACC, and absent cingulate gyrus (A). Axial MRI showing Probst bundle (arrow, B).



Figure 3: Sagittal MRI demonstrating a suspicious persistent primitive olfactory artery (arrow) and definite anterior commissure (arrowhead).



Figure 4: CT angiography demonstrating right persistent POA (arrow) (A). CT angiography demonstrating a long anterior communicating artery (B, arrowhead).



Figure 5: CT angiography sagittal image demonstrating a highly located internal cerebral vein (arrow).

is complete in its craniocaudal extent by 18 to 19 weeks, although further maturation and growth continues into postnatal life [1]. Early disruption in the maturation of the corpus callosum can lead to a developmental condition known as ACC, a relatively frequent congenital malformation ranging from complete absence to hypogenesis of corpus callosum fibers. This condition occurs in 1:4000 individuals, which makes it one of the most common human brain malformations [7]. The etiology of ACC has an identifiable cause in about 25% of cases [7], and is usually related to other neurological conditions such as hydrocephalus [8], microcephaly [9], or fetal alcohol syndrome [10].

However, the etiology of ACC is typically unknown. Because ACC is a heterogeneous condition, symptoms can vary greatly between affected individuals, ranging from relative absence to severe impairment requiring special education and assistance in everyday living [11,12].

The anterior commissure appears in the human fetal brain at a stage when the crown-rump length (CRL) is 31 mm (or approximately 7–8 weeks EGA), according to Müller and O’Rahilly [13]. Therefore, the development of the anterior commissure seems to occur earlier than the stage in which the first axon transverses the corpus callosum [14]. Cesaretti et al. [15], reported four groups of fetuses with ACC based on the presence or absence of other forebrain commissures as follows: Group 1 showed no forebrain commissure; Group 2 showed only anterior commissure; Group 3 showed anterior commissure and vestigial hippocampal commissure; and Group 4 showed anterior commissure and a hybrid structure composed of a vestigial hippocampal commissure and an early rudiment of the corpus callosum only. The present case falls into Group 2 ACC with anterior commissure only. ACC associated with intact anterior commissure may result from an event occurring around 7–8 weeks EGA.

Developmental mechanism of persistent POA

Embryologically, early brain vesicles are supplied by a primitive internal carotid artery, which is further divided into rostral and caudal sections. The rostral division supplies the prosencephalon (rhinencephalon in lower vertebrates; thus, the artery is called the olfactory artery), and constitutes the POA. The POA terminates in the nasal fossa, and the secondary artery constitutes the medial olfactory artery, which supplies the olfactory bulb. The POA develops at the embryo developmental stage of 7–12 mm CRL at approximately 32–37 days EGA. The medial olfactory artery becomes the ACA proper, while the terminal portion of the POA typically regresses. Lateral olfactory branches of the POA include the recurrent artery of Heubner, anterior choroidal artery, lateral striate artery, and later middle cerebral artery [16]. At the embryo developmental stage of 18–24 mm CRL at approximately 43–48-days EGA, the ACA supplies the mesial surface of the cerebral hemispheres in the now definitive interhemispheric fissure [17].

We consider that persistent POA may develop from an event occurring after 37 days EGA. This event (mechanical or other factor) may result in no regression of the terminal portion of the POA and no development of the proximal ACA.

Brain vascular anomaly in a patient with ACC

Although no particular vascular abnormalities were found in rhinencephalic brains, no complete circle of Willis was present at the base of holoprosencephalic brains; the anterior part was lacking and replaced by anterior branches that emerged unilaterally or bilaterally from the internal carotid artery. The choroidal arteries were of very large caliber and ran to the highly vascularized wall of the dorsal cyst, which is usually present in holoprosencephalic brains [18]. van Overbeeke et al. [18], proposed that the vascular pattern of holoprosencephalic brains fits well into the embryological concept of the dominance of neural over vascular development. This may support the concept of the role of functional demand.

Cerebral vascular anomalies associated with ACC are sinus pericranii [3], meandering of the ACA, coursing of the ACA directly upwards in an interhemispheric fissure, and persistent falcine sinus [4]. According to van Overbeeke et al. [18], neural development is dominant to vascular development. Although patients with ACC may have sensory and visual deficits, these patients are surprisingly functional in everyday living. We consider that ACC results in less severe cerebrovascular anomalies that are mostly insignificant from a clinical standpoint.

To the best of our knowledge, this is the first reported case of ACC associated with persistent POA. We consider that ACC associated with persistent POA may result from some event during the embryological period. The typical findings in this case were as follows: total ACC, intact anterior commissure, and persistent POA. Therefore, this event may have occurred between 32–37 days and 8 weeks EGA.

Conclusion

To the best of our knowledge, this is the first reported case of persistent POA associated with ACC. Development of the corpus callosum and POA are closely related in developmental time. We consider that one event between 32–37 days and 8 weeks EGA may result in persistent POA, intact anterior commissure, and ACC. The findings in this case suggest that CT angiography or MRI may detect persistent POA more effectively in cases involving ACC.

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