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Case Report

UNILATERAL COLPOCEPHALY WITH BILATERAL PARTIAL AGENESIS OF CORPUS CALLOSUM IN ADULT MALE: A CASE REPORT

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ABSTRACT

Colpocephaly, a rare congenital brain abnormality characterized by disproportionate enlargement of the posterior horn of the lateral ventricles, typically presents with neurological deficits in infancy. We present a unique case discovered incidentally during routine cadaveric dissection of a 63-year-old male with no apparent symptoms. The brain exhibited massive colpocephaly, accompanied by bilateral partial agenesis of the corpus callosum.

Morphometric analysis revealed significant enlargement of the left lateral ventricle, with an anterior-to-posterior ratio indicative of colpocephaly rather than normal pressure hydrocephalus. Comparative studies with non-colpocephalic brains corroborated these findings. Literature review suggests a developmental origin for colpocephaly, possibly related to disturbances in hydrostatic balance during embryogenesis.

Previous reported cases of adult-onset colpocephaly underscore its rarity and diagnostic challenges. Understanding the distinctive morphometry of colpocephalic lateral ventricles is crucial for accurate diagnosis, especially to differentiate it from more common conditions like idiopathic normal pressure hydrocephalus, thus avoiding unnecessary interventions. This case highlights the importance of cadaveric studies in uncovering rare anatomical variations and expanding our knowledge of neurological conditions.

Keywords : Colpocephaly, Morphometric analysis, Corpus callosum agenesis

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INTRODUCTION

Colpocephaly is a rare congenital abnormality in the ventricular system of the brain. It is characterized by disproportionate enlargement of the posterior horn of the lateral ventricle as compared to the anterior horn. Other neurological malformations, particularly agenesis of the corpus callosum and microgyria, are associated with this condition [1]. It can be differentiated from hydrocephalus, where there is proportionate ventriculomegaly of the horns of the lateral ventricles [1]. The radiological diagnosis is usually made in the prenatal period and later manifests as intellectual disability [2].

CASE REPORT

In routine dissection for undergraduates at Government Doon Medical College, Dehradun, Uttarakhand, King George Medical College, Lucknow, and CDSIMER, Harohalli, Ramnagara district from the period of 2020 to 2023, a total of 20 sagittal sections of brains were examined. During brain removal after removing the calvaria, a striking case of massive colpocephaly with small cerebral white matter in the parietal and occipital lobes of the left cerebral hemisphere, along with bilateral partial agenesis of the corpus callosum, was incidentally reported in a 63-year-old male cadaver who was otherwise a functional male and had a natural death at CDSIMER, Harohalli, Ramnagara district in 2021, as reported by his death certificate. The rest of the 29 brains were found to have normal anatomy of lateral ventricles. To our best knowledge, this is the first cadaveric reported case in an asymptomatic adult and is described in the cadaveric literature.

Morphometric analysis of the colpocephalic sagittal section of the brain was conducted using an inch tape. Various parameters such as anteroposterior length of the anterior and posterior horns and body of the lateral ventricle, and height (superoinferior length) of the body of the left ventricle were measured with an inch tape in sagittal and transverse sections of the brain. The posterior-to-anterior (P/A) ratio was also calculated by taking the maximal width of the posterior horn and dividing it by the maximal width of the anterior horn of the lateral ventricle in the transverse section of the left cerebral hemisphere.

RESULTS

The right cerebral hemisphere of this cadaveric male seems to have had normal anatomy except for partial agenesis in the center of the body of the lateral ventricle. However, in the left cerebral hemisphere, massive disproportionate enlargement of the lateral ventricle was observed, with an anterior-posterior length of 13.2 cm and superoinferior length (height) of 6.5 cm, resulting in a P/A Ratio of 3.1 (>3). The maximal width of the posterior and anterior horn was found to be 4.96 cm and 1.6 cm, respectively, which highly suggests the case of colpocephaly rather than normal pressure hydrocephalus [3]. (Figs. 1-4)

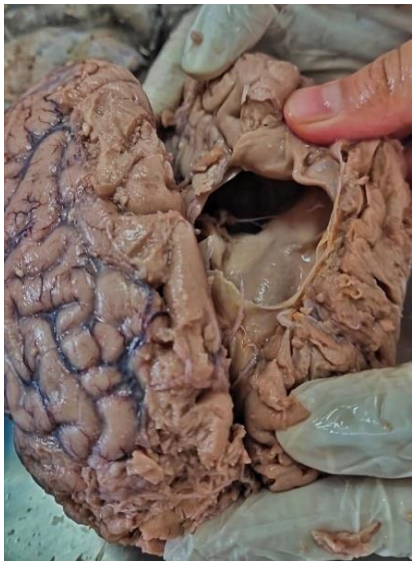

Fig. 1. Case of colpocephalic brain (intact right and left cerebral hemisphere) in routine dissection

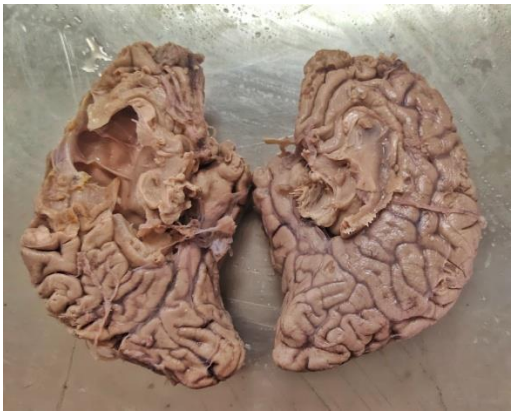

Fig. 2. left and right cerebral hemisphere showing Massive colpocephaly on left side and bilateral partial agenesis of corpus callosum on right side

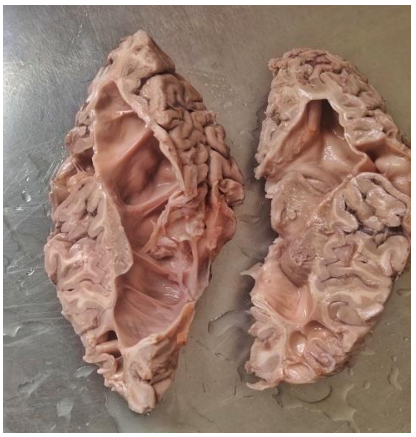

Fig. 3. Transverse section of left cerebral hemisphere showing massive colpocephaly showing the maximum width of posterior and anterior horn measurement

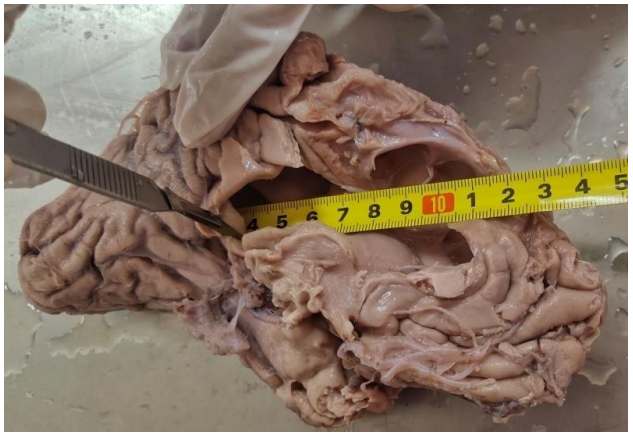

Fig. 4. Measurement of length of posterior horn of left cerebral hemisphere

DISCUSSION

Colpocephaly is typically discovered in infancy due to associated intellectual disability, seizures, motor abnormalities, or visual abnormalities [4]. Discovery in adulthood is remarkably uncommon. Colpocephaly can be identified radiographically by measuring the maximal width of the anterior and occipital horns of the lateral ventricles. An occipital-to-anterior horn ratio of greater than 3 is highly specific for colpocephaly, although it has relatively low sensitivity [5].

According to Benda et al.'s study, intermediate zone fibers originating from the thalamus and corpus callosum in vesiculocephaly fail to develop at the end of the fifth embryonic month, and its architectonic appearance suggests colpocephaly [6]. Yakovlev et al. stated that colpocephaly results from a disturbance in the hydrostatic balance between the intra- and extraventricular pressures as a consequence of failure of the development of the cerebral wall, resulting in outpocketing of the ventricular wall. Their study also stated that other anomalies seen in association with colpocephaly suggest an insult not later than the second embryonic month [7].

Noorani et al. in their CT study of 14 cases of colpocephaly in a series of 3,411 consecutive CT scans in the 1988 California population found the P/A ratio as 2.90±1.44, which almost approximates our findings. This study also stated that periventricular leukomalacia, which results from the destruction of the optic radiations and subsequent degeneration in the white matter of the occipital lobe, could be the possible explanation for the development of disproportionately enlargement of the occipital horns of the lateral ventricles [8].

Honnegowda Tm et al. reported a mean anteroposterior length of the body and frontal horn of the lateral ventricle in non-colpocephalic brains as 7.6 cm and 3.0 cm, respectively, in their CT study of the brain, which is close to our findings, thereby proving that in the present study, the defect was present only in the posterior horn, suggesting more of colpocephaly [1].

C Gyldensted et al. found the width of the left anterior horn as 1.9 cm in the CT study of non-colpocephalic brains, which approximates our finding, thereby proving that no abnormality is present in the anterior horn. Therefore, it proves our hypothesis that the present case was of colpocephaly, which does not involve anterior horns [9].

Duffner F et al. in their MRI study for non-colpocephalic brains, found the mean total length (anteroposterior) of the lateral ventricle, posterior horn, and height of the lateral ventricle as 9.1 cm, 2.8 cm, and 1.7 cm, respectively, which are not found to be in unison with our findings (13.2, 4.2 cm, and 6.5

cm respectively), suggesting that in our present case of the brain, dimensions found were abnormal, proving the hypothesis of colpocephalic brain [10].

Previously reported cases of colpocephaly were diagnosed incidentally in adulthood during CT examination post-onset of mild neural symptoms in Wunderlich G, et al. in 1996 [11], Cheong J, et al. in 2012 [12], Esenwa C, et al. in 2013 [13], Brescian N, et al. [14], Nasrat T, et al. in 2014 [15], Bartolome E, et al. in 2016 [16].

CONCLUSION

Colpocephaly discovered in asymptomatic adults is exceedingly rare. It may be misdiagnosed as normal pressure hydrocephalus. Knowing the respective morphometry of the lateral ventricle can aid clinicians in differentiating this disproportionate ventriculomegaly affecting the posterior horn and body of the left lateral ventricle in a colpocephalic brain from that of the common form of adult ventriculomegaly (idiopathic normal pressure hydrocephalus) and thereby preventing unnecessary interventions.

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