

Colpocephaly and Corpus Callosum Agenesis in an Asymptomatic Adult

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A sixty-nine-year-old Bahraini female, known case of type II diabetes mellitus (DM), hyperlipidemia and hypertension (HTN) presented with a history of acute left-sided lip numbness and right-sided mouth deviation. The neurological manifestation started suddenly and progressed gradually.

Computerized tomography (CT) and magnetic resonance imaging (MRI) of the brain revealed absent organic causes for the neurological manifestation; however, there was disproportionate dilatation of the occipital horns of lateral ventricles and corpus callosum agenesis. The findings were suggestive of colpocephaly.

Colpocephaly is a condition known to affect children. It leads to seizures and gait abnormality. It is almost always diagnosed in childhood. We reported this case due to its rare presentation in old age and the absence of neurological deficit throughout the patient's life.

Bahrain Med Bull 2019; 41(4): 275 - 277

The brain ventricles form communication network cavities that have cerebrospinal fluid (CSF) and are lined with ependymal cells. The ventricular system is located in the brain parenchyma and has two lateral ventricles, the third ventricle forms the cerebral aqueduct and the fourth ventricle is situated in the brain stem¹. The cerebral spinal fluid is mostly but not exclusively, secreted by the choroid plexus. The brain interstitial fluid, ependymal cells and capillaries play a poorly understood role in the cerebral spine fluid secretion. The choroid plexus secrete the cerebral spinal fluid at a rate of 0.2-0.7ml per minute or 600-700ml per day. The mean cerebral spine fluid volume is 150ml; 25ml of those are located in the ventricles and 125ml are found in the subarachnoid spaces².

Colpocephaly is a congenital abnormality of the ventricular system of the brain that is recognized in pediatric age and could present as motor system abnormalities, intellectual disabilities, visual defects, and speech and language difficulties³. The condition leads to the disproportional enlargement of the occipital horns of the lateral ventricles. It could be due to heterogeneous disorders of the neuroblasts during early development, central nervous disorders, perinatal injuries or intrauterine disturbances. In addition, it is believed that maternal exposure to corticosteroids during pregnancy, toxins and toxoplasmosis infections lead to the development of chromosomal anomalies and perinatal anoxic-ischemic encephalopathy.

The condition is diagnosed by CT and MRI, which reveal the cephalic abnormalities and enlargement of the occipital horns of the lateral ventricles. There are no definitive treatments for

patients, and anticonvulsant medications are prescribed to prevent seizures.

The aim of this report is to present a case of colpocephaly found in an adult patient and the patient was neurologically asymptomatic throughout her life.

THE CASE

A sixty-nine-year-old Bahraini female, known case of type II DM, hyperlipidemia and HTN presented with a history of acute left-sided lip numbness and right-sided mouth deviation. The neurological manifestation started suddenly and progressed gradually. The patient had a similar episode four years earlier on the same side. However, prior to that, she has been asymptomatic and did not complain of any neurological symptoms. The patient denied any gait instability or seizure. The patient was alert, conscious and oriented with a Glasgow Coma scale of 15/15. She was unable to close her left eye tightly; the mouth deviated to the right. No wrinkles were found on the left side of the face. Reflexes were normal, power 5/5 on all limbs, no absent or lost sensations. Other systems were unremarkable.

CT brain revealed dilated occipital horn of lateral ventricles, such finding is suggestive of colpocephaly see figure 1.

MRI of the brain confirmed the CT findings and the absence of corpus callosum, which confirmed the diagnosis of colpocephaly. No additional organic causes were detected, see figures 2 and 3.

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Figure 1: Axial Non-Enhanced CT of the Brain Showing Disproportionate Dilatation of the Occipital Horns (White Arrows) in Comparison to the Frontal Horns (Yellow Arrows) of Lateral Ventricles

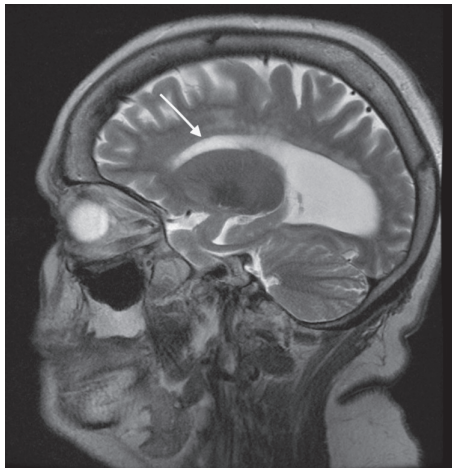


Figure 2: Mid-Sagittal View T2WI MRI Sequence Showed Absent Corpus Callosum and Only Cingulate Gyrus (White Arrow) Seen Overlying the Third Ventricle

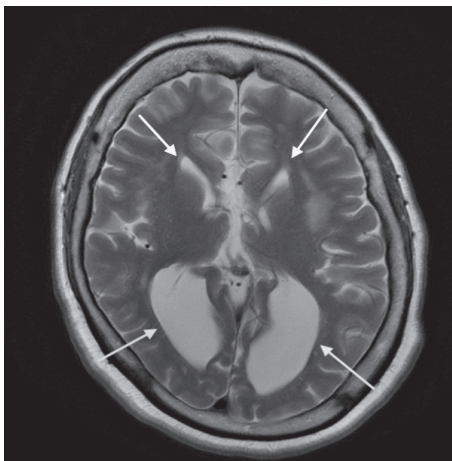


Figure 3: Axial, T2W Sequence MRI Showing Disproportionate Dilatation of the Occipital Horns (Yellow Arrows) of Lateral Ventricle in Comparison to Frontal Horns (White Arrows)

DISCUSSION

Srivastava et al found that there were limited numbers of colpocephaly cases discovered in adulthood⁴. Esenwa et al reported a case of adult colpocephaly where the patient was symptomatic throughout her life³.

Our patient has been asymptomatic throughout her life; her cross-sectional findings were confirmatory of colpocephaly. Therefore, colpocephaly should be considered in the differential diagnosis even if the patient is asymptomatic adult if the cross-sectional imaging revealed disproportionate ventricular dilation of the occipital horns of the lateral ventricle, rather than labeling the patient as normal pressure hydrocephalus because both diagnoses have different management approaches.

Antenatal diagnoses that are based on the non-visualization of the corpus callosum are infrequent⁵. In our case, the elderly patient was diagnosed by CT scan of the brain.

Symptoms of colpocephaly in children include seizures, mental retardation, poor vision and speech and language difficulties⁶. MRI or CT scans of the children with colpocephaly show marked enlargement of the occipital horns of the lateral ventricles and dilated ventricles with partial or full agenesis of the corpus callosum. A differential diagnosis that rules out normal pressure hydrocephalus in adult colpocephaly patients should be considered before brain shunting is performed⁷.

Normal-pressure hydrocephalus (NPH) is an adult-onset form of progressive ventriculomegaly that results from the buildup of the CSF in the ventricles due to the blocking of the normal CSF flow in the brain and spinal cord. The accumulation of CSF in the ventricles leads to enlargement and an increased pressure on the brain⁸. Normal-pressure hydrocephalus leads to mental impairment and dementia and shunting leads to improved patient outcomes. The differential diagnosis of the two is by CT and MRI. Colpocephaly is diagnosed by the enlargement of the occipital horns that result in a posterior to anterior ratio of ≥ 3 , while normal pressure hydrocephalus is diagnosed by the clinical triad: gait, cognitive and urinary dysfunction⁹. A posterior-to-anterior ratio of ≥ 3 is specific to colpocephaly, the configuration of the lateral ventricles, history and physical findings of the patients could be used to differentiate the cephalic disorders. MRI measures aqueductal flow rate. Evans and intracranial compliance indexes and callosal angle could be used to distinguish between colpocephaly and normal pressure hydrocephalus¹⁰. The Evans index shows the ratio of maximum width of the frontal horns of the lateral ventricles to the maximum internal diameter of the skull and a ratio of >3 indicate normal pressure hydrocephalus¹¹.

Hydrocephalus could be either communicating or non-communicating. The non-communicating hydrocephalus could be caused by colloid cysts, congenital aqueductal stenosis, masses, hematoma or infarcts. The colloid cysts are one of the classic causes of obstruction at the level of the foramina of Monro. Congenital aqueductal stenosis is a common X-linked recessive disorder that is seen in early childhood. Masses of the pineal gland are the most common cause of extrinsic obstruction of the aqueduct. The communicating hydrocephalus could be caused by infection, hemorrhage, and tumors. The arachnoid

villi are delicate structures that might be affected by insults, which lead to communicating hydrocephalus¹².

Our patient had been free of neurological complaints all her life, and the diagnosis was an incidental finding on CT. The radiological findings showed agenesis of the corpus callosum, non-obstructed ventriculomegaly and uneven dilation of the occipital horns.

CONCLUSION

The differential diagnosis of ventricular dilatation in adults include brain age related process with cerebral atrophy and hydrocephalic changes; both could be excluded by the presence of isolated dilation of the posterior horns of the lateral ventricles with normal diameter of the anterior horns. The latter findings are suggestive of colpocephaly.

Author Contribution: All authors share equal effort contribution towards (1) substantial contributions to conception and design, acquisition, analysis and interpretation of data; (2) drafting the article and revising it critically for important intellectual content; and (3) final approval of the manuscript version to be published. Yes.

Potential Conflicts of Interest: None.

Competing Interest: None.

Sponsorship: None.

Acceptance Date: 21 August 2019.

Ethical Approval: Approved by the Ethical Committee, Bahrain Defence Force Hospital, Bahrain.

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