

Obsessive Patient with Agenesis of the Corpus Callosum

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Abstract

Agenesis of the corpus callosum is an uncommon congenital anomaly. The following is a report of a patient with agenesis of the corpus callosum with cavum septum pellucidum and colpocephaly. This patient has also been diagnosed with anankastic personality disorder.

Keywords: *Corpus callosum, Septum pellucidum, Personality disorders*

Introduction

The corpus callosum is the largest commissural connection between the 2 cerebral hemispheres. Anatomically, the corpus callosum is inferior to the longitudinal fissure and superior to the diencephalon. It is divided into 4 parts: the rostrum, body, genu, and splenium. Tumour, infarct, or injury to the corpus callosum can lead to interhemispheric transfer deficit, depending on the site and extent of involvement.¹ The so-called 'disconnection syndrome' was demonstrated by Sperry in 1974, in which split brain individuals whose corpus callosum was surgically sectioned were recruited for split brain experiments.² The experiment brought greater understanding of hemispheric function.

In psychiatric practice, agenesis of the corpus callosum (ACC) is an uncommon congenital malformation. It is usually detected in childhood, whereas in adults it is paucisymptomatic.³ ACC accounts for 14% of central nervous system malformations.⁴ Some patients with ACC are asymptomatic and ACC is only detected incidentally by neuroimaging such as computed tomography (CT) of the brain or magnetic resonance imaging (MRI). Nevertheless, some patients may present to a psychiatrist because of epileptic seizures,⁴ mental retardation, developmental delay, or a psychotic experience. However, there is usually no significant relationship between ACC and psychosis, although some studies have suggested an association between posterior fossa atrophy or cerebellar anomalies and schizophrenia-like psychosis.⁵ ACC can be a discrete and isolated entity

but, occasionally, it is associated with Dandy-Walker cysts, hydrocephalus, meningomyelocele, microgyria, arachnoid cysts, interhemispheric cysts, porencephaly, Arnold-Chiari II malformations, skull deformities, facial dysmorphism, in-born errors of metabolism, neurocutaneous disease, or chromosomal microdeletion or aberrations^{4,6-9} Associations with ACC have also been found with the following illnesses — asplenia syndrome, multiple congenital anomaly syndrome, X-linked mental retardation syndrome, Andermann syndrome, DiGeorge-velocardiofacial syndrome, Aicardi syndrome, and Coffin-Lowry syndrome.^{5,8,10-13} X-linked recessive inheritance¹⁴ and autosomal recessive inheritance¹⁵ have been most commonly reported.

The prognosis depends on the presence of cerebral malformations that are involved in epilepsy and cognitive disturbances. There was a case report of a patient with ACC with strong anatomical and functional asymmetries.¹⁶ Strong right hand superiority for hand skill and tactile object recognition indicated unusual left hemispheric dominance for these functions.¹⁶ On the other hand, research has revealed that patients with ACC do not have disconnection syndrome as do individuals with split brain. Symptoms of interhemispheric disconnection are typically less severe for patients with ACC than after surgical section of the corpus callosum. It has been suggested that functional compensation is less efficient if ACC is partial rather than complete.¹⁷ However, ACC is associated with specific problems in complex cognitive operations such as tests on reasoning and concept formation. In patients with ACC, significantly poor performance has been found for tests of social insight, proverb interpretation, social logic, self-perception, and interpretation of ambiguous stimuli.¹⁸ The following is a report of an adult man with complete ACC, cavum septum pellucidum, colpocephaly, and concomitant anankastic personality disorder.

Case Report

A 34-year-old unmarried man, living with his parents, was referred to the South Kwai Chung Psychiatric Centre from the Medical Unit of the Princess Margaret Hospital due of worsening of work and social performance. He had an

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unstable work record due to poor output. He was slow to learn and had difficulty memorising material. He was slow and had difficulty in following instructions. Ongoing relationship problems with other people were also reported.

The patient's first attendance at the South Kwai Chung Psychiatric Centre was in early October 2000. There was no family history of mental illness, epilepsy, or genetic disease. He was born in Hong Kong and was the fifth born of 7 siblings. His birth and childhood was uneventful and no delay in developmental milestones was reported. The patient had a low average academic performance at school. He repeated form 5 and passed all his HKCEE exams at the second attempt. Later, he studied at a university in Canada and graduated with an average degree. He returned to Hong Kong and lived with his parents.

This patient had an unstable job record. He tried working at various jobs, but could only stay for 3 months in each of them. The longest job he had lasted for 5 years. His last job only lasted for a few weeks. The patient was inadequate, introverted, and prone to low self esteem. He was avoidant, dependent, and passive, and tended to hide his feelings. He had few friends and the only intimate relationship he had was 10 years previously, which lasted for only 3 months. He did not get along with his siblings apart from his immediate elder sister. He had no hobbies and was obsessed about minor details and could not tolerate minor mistakes. He was obstinate about doing things his own way and tended to read and learn the details before starting a project. He was unrealistic in goal setting and lacked emotional reactivity to external events. He had never smoked and had no history of substance or alcohol abuse. His past health was unremarkable. There was no history of epilepsy, head injury, operation, or drug allergy.

Mental State Examination

Mental State Examination revealed that the patient had no dysmorphic features. He dressed appropriately and took reasonable care of himself. He appeared cold and distant, although he could maintain a certain amount of eye contact. His speech was fast, relevant, and coherent but over-elaborated. His mood was euthymic while his affect was restricted. He did not harbour any suicidal ideas. His sleep and appetite were normal. No psychotic symptoms could be elicited. No overt rumination or compulsive ritual was present. He was orientated in time, place, and person. Immediate registration, 5-minute recall, and long-term memory were all normal. His intelligence was average and he obtained an intelligence quotient score of 95. His memory function as indicated by the Wechsler Memory Scale-Revised¹⁹ was of low average standard. He had specific problems with the recall of visual memory, especially complex visual stimulation. His attention and concentration was of average to above-average standard. His memory function was below expectation given his education level. He was poor in proverb interpretation and reasoning. He agreed to have further follow-up for insight but did not consider himself to have any mental illness.

Physical Examination

The patient's general physical condition was good. His cardiovascular system, chest, and abdomen were essentially normal. Neurologically, he was found to have normal gait and intact cranial nerve functions. His sensory, motor, and proprioceptive functions were all preserved. No frontal lobe signs were elicited. He was tested specifically for functional disconnection of cerebral hemispheres. The patient was blindfolded and objects were put into his hands. He was asked to name them one by one. Arabic numbers and letters of the alphabet were written on his palms and he was asked to read them aloud. Verbal commands were made to both ears to perform certain gestures. The results were all negative. There was neither callosal left-sided apraxia nor astereognosis. The patient is right-handed.

Assessment by Occupational Therapist

In a work capacity evaluation by standardised assessments of Valpar Components Work Sample 6 — Problem Solving conducted by an occupational therapist, the patient achieved a methods-time measurement standard at the 130 percentile for the work sample. This indicated that he attained the entrance level for the tested aptitudes and might exceed the level. For the Employee Attitude Survey, the patient achieved a range from the 50 to 99 percentile indicating that he was almost competent for working as a 'junior clerk' in open employment. During the test, the patient frequently asked for an explanation of the assessment details in order to avoid making mistakes. He had no problem in receiving instructions and was able to follow the assessment items accordingly.

Investigations

Basic laboratory results including complete blood cell count, erythrocyte sedimentation rate, and liver and renal function tests were normal. Electroencephalogram showed transient spikes in the left temporal region. Otherwise, it was composed of posterior prominent alpha activity admixed with a moderate amount of anterior prominent beta activity. No definite pathology could be identified. CT of the brain showed that "Lateral ventricles appeared parallel to each other and third ventricle was high riding. The posterior part of the corpus callosum was not visible and the anterior part was not well seen. A 1 cm x 2.5 cm hypodense, non-enhancing structure was seen on the left lateral aspect of the third ventricle anteriorly. There was no contrast enhancement. There was no midline shift or extra-axial collection." The impression was that there was complete or partial absence of the corpus callosum and a hypodense lesion near the third ventricle. MRI showed that "Complete agenesis of corpus callosum was noted. There was high riding third ventricle and separation of the lateral ventricles. Colpocephaly was also noted. The lesion on the left lateral aspect of third ventricle was still seen. It was of the same signal as cerebrospinal fluid in all sequences. No contrast enhancement was seen." The impression was that there was complete corpus callosum agenesis with a cystic structure on the left lateral aspect

of the third ventricle (cavum septum pellucidum). The brainstem auditory evoked potential showed normal form and latency without lateralisation features. Pattern-reversal visual evoked potential showed delayed P100 for both left and right eyes. Flash visual evoked potential delay was significant only for the right eye. The left eye potential was satisfactory.

Diagnosis

An Axis II diagnosis of anankastic personality disorder was made according to International Classification of Diseases, 10th edition, criteria. A physical diagnosis of ACC, colpocephaly plus cavum septum pellucidum was also established.

Discussion

Although no formal cognitive tests were done, there were explicit cognitive and psychosocial deficits in this index patient. Such phenomena could be related to diminished interhemispheric transfer. This picture was compatible with other case reports.¹⁸ Abnormalities or ACC have been reported in relation to schizophrenia in many studies.²⁰⁻²⁴ It has even been suggested that schizophrenia is a split-brain condition akin to ACC, but unrecognised due to the use of compensatory ipsilateral sensory pathways. Abnormalities of the septum pellucidum have also been found to be associated with schizophrenic illness.²⁵⁻²⁷ Agenesis of the corpus callosum, however, is not known to be associated with personality disorder. This paper reports a patient with ACC, cavum septum pellucidum, and colpocephaly. The patient also showed features of anankastic (obsessional) personality disorder.

In the literature, abnormality in the corpus callosal signal intensity in treatment-naïve obsessive compulsive disorder (OCD) has been reported, particularly in the region connecting the ventral prefrontal cortex and the striatum. Signal intensity is a reliable index of myelinisation of the corpus callosum.²⁸ In another study, it was found that all the regions of the corpus callosal except the isthmus were significantly larger in patients with OCD than in controls. The corpus callosal area correlated significantly with OCD symptom severity but not illness duration.²⁹ Nevertheless, OCD is nosologically different from anankastic personality disorder, although there may be a certain aetiological relationship. On the other hand, colpocephaly is an abnormal congenital enlargement of the occipital horns of the lateral ventricles. It is also described as persistence of the foetal configuration of the lateral ventricles into postnatal life. It has a typical association with learning disability (moderate to severe), seizures, and motor and visual abnormalities. These were not found in this patient. Colpocephaly may coexist with absence of the corpus callosum and cavum septum pellucidum,³⁰ in which agenesis of the corpus callosum is the most frequently associated malformation. Colpocephaly is a marker of disordered brain formation and an error in morphogenesis of diverse origins. Encephaloclastic insults occurring at any

time between 1 and 4 months gestation may result in this anomaly. In addition, colpocephaly may have associated chromosomal anomaly. Unfortunately, this patient refused genetic study.

Conclusion

To conclude, this is the first report of anankastic personality disorder in a patient with ACC with coexistence of cavum septum pellucidum and colpocephaly. The relationship between ACC and various personality disorders may need further investigation and research. Nonetheless, the presence of anankastic personality disorder in an index case may just be a coincidence.

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