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Arnold-Chiari malformation and agenesis of the corpus callosum in a case of brief psychotic disorder

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Summary

We report a case of a brief psychotic disorder with type I Arnold-Chiari malformation and agenesis of the corpus callosum which was detected on a magnetic resonance imaging (MRI) scan. The patient was started on antipsychotics and underwent decompression surgery. He is on regular follow-up and returned to normal daily life within a few weeks. The antipsychotics were tapered and eventually stopped, and the patient is maintaining well in the follow-up. With improved neuroimaging techniques reporting brain morphological abnormalities in psychiatric patients has increased and we suggest that a neuroimaging investigation is valuable in psychotic illness.

Arnold-Chiari malformation, agenesis of the corpus callosum, brief psychotic disorder

INTRODUCTION

Arnold-Chiari malformations (ACM) are a group of related congenital anomalies at the base of the brain. ACM is a very rare disorder with displacement of the cerebellar tonsils caudally into the foramen magnum. To date, the cause of this malformation is unknown; however, there is some evidence that a defect of the paraxial mesoderm plays a part, as it results in a shallow posterior cranial fossa and brainstem as well as herniation through the foramen [1].

The corpus callosum (CC) is the largest white matter tract in the brain. It connects most bilateral homologous regions of the cerebral cortex and carries information between them [2,3]. It is made up mostly of glutamatergic fibers, which become progressively myelinated with age. The development of the CC is associated with the formation of the hippocampus and cingulate cortex, which are implicated in the pathophysiology of schizophrenia. However, whether the CC is abnormal in patients with schizophrenia is unclear [4,5]. The incidence of agenesis is difficult to estimate because the anomaly is rare and not routinely detected [6].

With the spread of neuroimaging diagnostic equipment and improvement in the resolution of achieved images, reports on morphological abnormalities of cerebral white matter among psychiatric patients are increasing. A number of magnetic resonance imaging (MRI) studies on the brains of patients with schizophrenia showed cortical and subcortical abnormalities, including in the corpus callosum. Reports of schizophrenic patients with partial or complete agenesis of the corpus callosum, which is associated with neurodevelopmental abnormalities, have occasionally been cited [7,8].

Here we report a case of a patient who presented with a brief psychotic disorder. Neuroim-

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aging detected type I Arnold-Chiari malformation with agenesis of the corpus callosum.

CASE REPORT

A male patient aged 22 who was working as a day laborer in a factory was brought by his father to the psychiatric clinic in Vydehi Institute of Medical Sciences and Research Centre with complains of abnormal behavior, fearfulness, suspiciousness, decreased sleep and appetite, and abstaining from work for the past 8 days. There was no history of head injury, substance use or fever; no history of prenatal trauma or exposure to drugs; and no developmental delay (his academic achievement was average). The patient had left schooling at the age of 14 due to financial constraints. There was no history of psychiatric illness, physical illness and no family history of any serious illness. On examination his vitals were normal and the systemic examination was normal too. Neurological examination revealed normal cranial nerves as well as normal sensory and motor systems. The patient was well oriented and had no memory problems. He had delusions of reference and persecution, and both visual and auditory hallucinations. A battery of investigations was performed (complete hemogram, blood sugar, renal function, liver function test, serum lipid profile, serum electrolytes, thyroid function test) and all were found to be within normal limits. Viral serology (HIV 1&2, VDRL) was non-reactive.

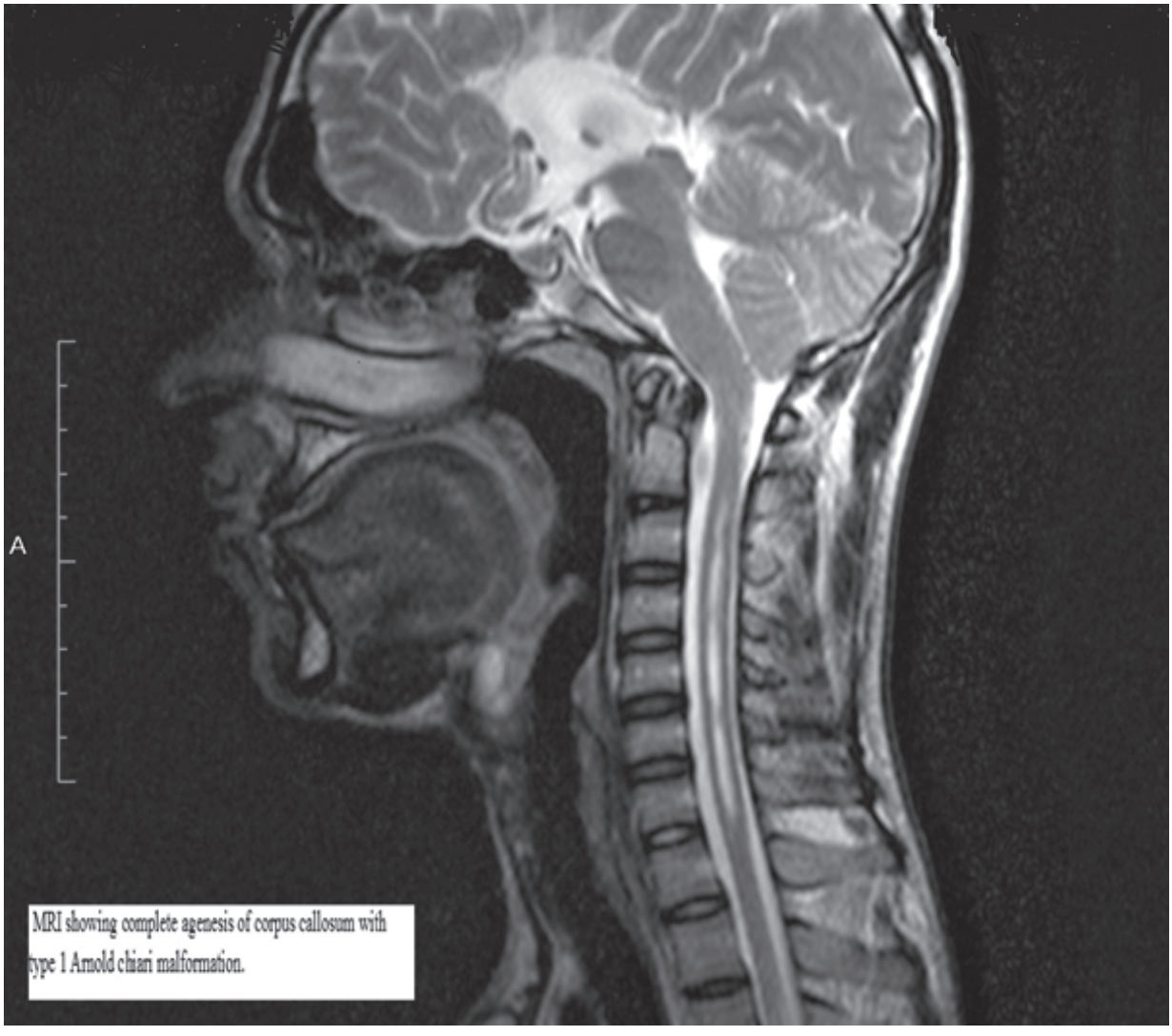

Fig. 1 MRI showing complete agenesis of corpus callosum with type 1 Arnold Chiari malformation.

However, an MRI brain scan showed a complete agenesis of the corpus callosum with type 1 Arnold-Chiari malformation.

The patient was started on olanzapine 10 mg once daily to control the psychotic symptoms. He was referred to a neurosurgeon and after two weeks, once the psychotic symptoms were controlled, underwent a foramen magnum decompression surgery. Following the surgery, his olanzapine was tapered down to 5 mg and over a period of 9 months stopped altogether. The patient reached premorbid functioning after 6 weeks and went back to work. He has reported for regular follow-up over the past year and has stayed well without medication.

DISCUSSION

ACM is characterized by four subtypes:

- Type I involves a herniation of the cerebellar tonsils into the foramen magnum.
- Type II is associated with a myelomeningocele and hydrocephalus, herniation not only of the tonsils but also the vermis, the fourth ventricle and the pons. Morphologically, aqueductal stenosis, hydromyelia and cortical dysplasia occur.
- Type III is characterized by an encephalocele, the descent of both the cerebellum and the brainstem into the spine and internal sac.
- Type IV is associated with cerebellar atrophy [1].

ACM is best diagnosed with MRI [1]. Its prevalence ranges from 0.2 to 1% [9]. There are many somatic complications caused by ACM such as pain, motor deficits, hand muscular atrophy, lower cranial palsy, cerebellar ataxia, nystagmus, sensory deficits, dysphagia and dysphonia [1]. In addition, psychiatric comorbidities such as anxiety and mood disorders occur and affect functioning and quality of life [10]. The neuropsychiatric symptoms in ACM are attributed to the compression of neuronal tissue, the nerves and blood vessels [11]. To date, only three case reports have described an association with psychosis: two cases were of type I ACM [11,12] and one of type II ACM [13].

Callosal deficit can be diagnosed by an MRI during the early developmental stage in utero or discovered because of various other anomalies during the postnatal development stage in the areas of pediatric neuroradiology and psychiatry [6].

A number of MRI studies on schizophrenic brains have shown cortical and subcortical abnormalities, including the corpus callosum. Volumetric decrease of the corpus callosum among schizophrenic patients is regarded as one of the reasons for cognitive dysfunction in the transfer of information between the two cerebral hemispheres. Schizophrenic patients with partial or complete agenesis of the corpus callosum, which is associated with neurodevelopmental abnormality, have been occasionally reported [7], as has delusional disorder with complete agenesis of the CC [14]. One review has suggested that abnormalities in the limbic structure developed along with corpus callosum agenesis may be linked to psychiatric disorder [15].

A damage or deficit in the corpus callosum is associated with cognitive dysfunction which could cause delusional symptoms transversely seen among specific psychiatric disorders. MRI investigations are valuable in the diagnosis of acute onset psychosis as they allow detecting any structural changes, whereas an integration of neuroimaging and cell biological studies is required to understand the correlations in pathogenesis between developmental failure and acquired brain damage [6].

In this case report, psychotic symptoms may be independently associated with ACM or agenesis of the corpus callosum or a combination of both, but as the psychotic symptoms have not resurfaced after decompression surgery we speculate that they were in fact attributed to ACM.

There are case reports as well as neuroimaging studies on the association of CC agenesis with schizophrenia and several case reports on the association of ACM and psychosis. So far, however, this is the first case report on the occurrence of both ACM and corpus callosum agenesis with psychosis. We would like to suggest that clinicians should be alert to organic disorders presenting with psychiatric symptoms.

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