Psychiatria Danubina, 2014; Vol. 26, No. 3, pp 291-293 *Case report*

PSYCHOTIC AND MAJOR NEUROCOGNITIVE DISORDER SECONDARY TO ARNOLD-CHIARI TYPE II MALFORMATION

Linda Hoederath, Lena Jellestad, Josef Jenewein & Soenke Boettger

Department of Consultation-Liaison Psychiatry, University Hospital Zurich, Zurich, Switzerland

received: 14.5.2014; revised: 14.8.2014; accepted: 23.8.2014

* * * * *

INTRODUCTION

Arnold-Chiari malformations (ACM) represent a constellation 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 for a defect of the paraxial mesoderm resulting in a shallow posterior cranial fossa and brainstem as well as herniation through the foramen magnum (Caldwell et al. 2009).

ACM is characterized by four subtypes. A) type I represents a herniation of the cerebellar tonsils into the foramen magnum, B) type II is associated with a myelomeningocele and hydrocephalus, herniation not only of the tonsils, but also the vermis, fourth ventricle, and pons. Morphologically, aqueductal stenosis, hydromelia and cortical dysplasia occur. C) type III is characterized by an encephalocele, the descent of both cerebellum and brainstem into the spine and internal sac, and D) type IV is associated with cerebellar atrophy (Caldwell et al. 2009).

There are many somatic complications caused by ACM such as a pain, motor deficits, hand muscular atrophy, lower cranial palsy, cerebellar ataxia, nystagmus, sensory deficits, dysphagia, and dysphonia (Caldwell et al. 2009). In addition, psychiatric comorbidities such as anxiety and mood disorders occur and affect the functioning and quality of life (Bakim et al. 2013, Mestres et al. 2012). To date, only two case reports describe the association with psychosis (Del Casale et al. 2012, Ilankovic et al. 2006) and only one case mentions a secondary major neurocognitive disorder (Mahgoub et al. 2012).

CASE REPORT

Mr B. is a 34-year-old Caucasian male with ACM type II, internal ventriculoperitoneal (VP) shunt, paraparesis of the lower extremity who was admitted to the burn-unit of the University Hospital Zurich after suffering from II-III degree burns of 21% of the body surface. He was wheelchair-bound, required substantial assistance and resided at a nursing home for the disabled. He had set himself deliberately on fire.

On admission, the patient was not able to interact due to his physical condition. Initial laboratory studies revealed an anemia and evidence for infection. The hemoglobin was 72 g/l, hematocrit 0.22 l/l, leukocytes were 15.9 G/l with 12.3 G/l neutrophiles. The C-reactive protein (CRP) was elevated with a value of 73. Electrolytes, liver function tests, thyroid function tests, and cerebral spinal fluid were all within normal limits. The ECG was normal and a cranial computed tomography (CT) scan confirmed the ACM type II with descended cerebellar tonsils, medullary kinking, and tectal beaking. The VP shunt was on the right, frontotemporally, the anterior horn of the left ventricle collapsed, the outer liquor space expanded due to the collapsed ventricular system, and a hypodense area right parietal-occipitally of unknown etiology noted.

Mr. B's psychiatric medications were lamotrigine 200mg daily, valproate 900mg daily and quetiapine 200mg daily.

During the course of management, the patient developed multiple infections, renal insufficiency and delirium which remitted. Once baseline was reached again, challenges in management arose due to his limitations and erratic behavior and a 1:1 was indicated throughout the course of hospitalization.

Past psychiatric history

Mr. B's. early years were unremarkable except for the physical limitations caused by the ACM. He was wheelchair-bound due to his lower extremity paresis, however, reached all milestones of development and was independent in the activities of daily living. His intellectual development was truly remarkable. He graduated from high school and qualified among the best for college.

However, in his adolescence the family noted episodes of being self-preoccupied, living detached in his own world, unable to interact indicating dissociation. In these states he displayed depersonalization - not being himself - and derealization - perceiving his environment in an altered way. The reality testing remained mostly intact, the symptoms caused significant distress and impairment in functioning, as well were not attributed to a substance or other medical condition and not attributed to another mental disorder.

At the age of eighteen, episodes of aggressive outbursts and uncontrollable behavior occurred, which the family primarily attributed to puberty. Then in his twenties, these behavioral disturbances developed into frank psychosis and the previous behavioral disturbances were identified as prodromal psychosis. Mr. B. showed erratic, disorganized behavior, had paranoid delusions of others pursuing and harassing him, wanting to steal from him and he was not able to distinguish whether the television set was talking to him or not. In addition, he heard various, imperative voices. He met three out of five DSM 5-criteria (APA 2013) for a psychotic disorder. Taking into account the severe pre-morbid brain pathology with multiple shunt complications which required revisions, recurrent episodes of hydrocephalus, the psychosis was attributed to the congenital brain anomaly and secondary complications. Overall, the patient was hospitalized more than thirty times, the hospitalizations added up to more than three years of his life. A long psychiatric career with eventually more than thirty hospitalizations, equaling more than three years in total and administration of most common typical and atypical antipsychotics, as well as mood stabilizers followed.

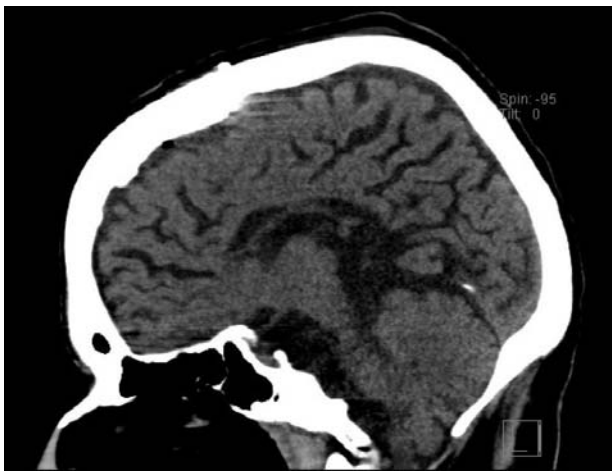

Figure 1. Sagittal cranial CT scan of ACM-II malformation

Mr B. continued to display depersonalization/derealization disorder episodes in which suicidal behavior became apparent such as attempting to access train tracks with his wheelchair or setting himself on fire. After these episodes ended, he usually felt guilty and shameful.

Although several attempts were made, the mother was able to handle the patient at home. For the last eight years he lived in various nursing homes and residences for the disabled, often being banned from them due to his erratic, uncontrollable, and self-endangering behavior. Within the last three years, his cognition and functional ability further declined. Progressive deficits in attention, concentration, executive function, memory, and apraxia evolved indicative of a major neurocognitive disorder. These deficits did not occur in the context of another mental illness, but were caused by the ACM-II, recurrent VP-shunt obstructions and episodes of hydrocephalus and met DSM-5 criteria for a major neuropsychiatric disorder secondary to this congenital brain anomaly. As a consequence, he required assistance in most activities of daily living. Most recently, the patient developed more severe dysarthria which made communication even more difficult.

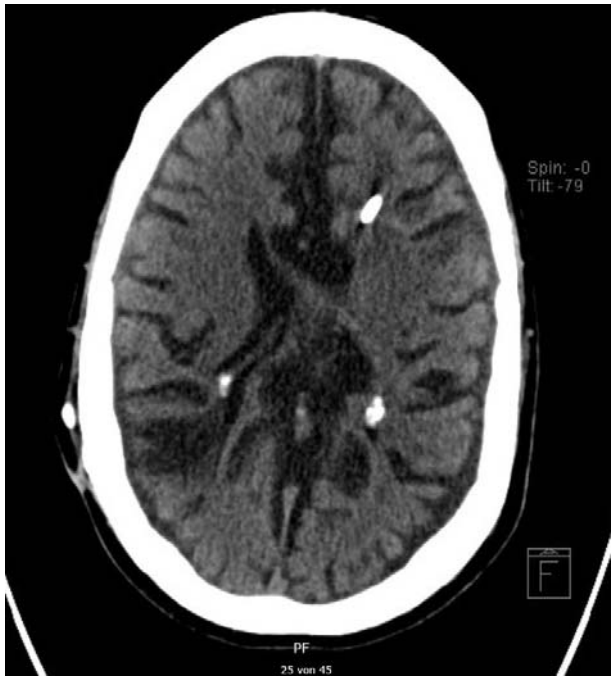

Figure 2. Transverse cranial CT scan of ACM-II malformation

DISCUSSION

This case represents several rare psychiatric comorbidities in the context of ACM-II. Although the functional and intellectual development was remarkable at first, a depersonalization-derealization disorder developed in the teenager years. Starting from the age of eighteen, a psychotic disorder and subsequently from the age of thirty on, a major neurocognitive disorder evolved.

Although age and presentation were characteristic of a schizophrenic illness, the severe cerebral malformation, ACM-II complicated by recurrent VP-shunt obstructions and episodes of hydrocephalus, classified this psychosis and subsequent major neurocognitive disorder as secondary to this medical condition. Within making differential diagnoses (First 2014), in the first two steps malingering or factitious disorder and substance-related etiologies had to be ruled out and were clearly not present in this case. In the next step, a direct effect of a general medical condition causing psychiatric symptoms had to be evaluated. From a differential diagnoses perspective this was the most difficult and challenging step as symptoms of a psychiatric disorder may be similar or identical to those caused by a medical

condition. In this case, the ACM-II, recurrent episodes of hydrocephalus secondary to VP-shunt obstructions preceded the onset of psychiatric symptoms. The course of psychiatric illness, at first presenting with depersonalization-derealization disorder, second with a psychotic disorder and at last with a major neurocognitive disorder represented an unusual course of psychiatric illness, which was not typical of a schizophrenic or major neurocognitive illness, in particular, taking into account the age at onset of the neurocognitive disorder. Thus, in particular the psychotic and major neurocognitive disorder encountered in this case, had to be attributed to ACM-II and its complicated course.

CONCLUSION

This is the first case report of an ACM-II anomaly with secondary psychotic and major neurocognitive disorder. So far, only two other cases of psychotic symptomatology (Del Casale et al. 2012, Ilankovic et al. 2006) and one case of dementia have been reported in the context of an ACM-I (Mahgoub et al. 2012). Among the cases with psychotic symptomatology, a psychosis risk syndrome with comorbid panic disorder was identified in the context of cannabis use and compression of the locus coeruleus (Del Casale et al. 2012) and recurrent psychotic episodes were attributed to epileptiform activities (Ilankovic et al. 2006). Thus, this is the first case of a psychotic and major neurocognitive disorder secondary to ACM- II complicated by recurrent VP-shunt obstructions and episodes of hydrocephalus.

Acknowledgements: None.

Conflict of interest: None to declare.

References

1. *American Psychiatric Association: Diagnostic and Statistical Manual of Mental Disorders (5th ed.). American Psychiatric Publishing, Arlington, VA, 2013.*
2. *Bakim B, Goksan YB, Yilmaz A, Karamustafalioglu O, Akbiyik M, Yayla S et al.: The quality of life and psychiatric morbidity in patients operated for Arnold-Chiari malformation type I. Int J Psychiatry Clin Pract 2013; 17:259-263.*
3. *Caldwell DL, Dubose CO, White TB: Chiari malformations. Radiol Technol 2009; 80:340MR-354MR.*
4. *Del Casale A, Serata D, Rapinesi C, Simonetti A, Tamorri SM, Comparelli A et al.: Psychosis risk syndrome comorbid with panic attack disorder in a cannabis-abusing patient affected by Arnold-Chiari malformation type I. Gen Hosp Psychiatry 2012; 34:702-707.*
5. *First MB: DSM-5 Handbook of Differential Diagnosis. American Psychiatric Publishing, Arlington, VA, 2014.*
6. *Ilankovic NN, Ilankovic AN, Bojic V, Ilankovic LM: Chiari I malformation in adults: epileptiform events and schizophrenia-like psychosis. Psychiatr Danub 2006; 18:92-96.*
7. *Mahgoub N, Avari J, Francois D: A case of Arnold-Chiari malformation associated with dementia. J Neuropsychiatry Clin Neurosci 2012; 24:E44-E45.*
8. *Mestres O, Poca MA, Solana E, Radoi A, Quintana M, Force E et al.: [Evaluation of the quality of life of patients with a Chiari type I malformation. A pilot study in a cohort of 67 patients]. Rev Neurol 2012; 55:148-156.*

Correspondence:
Linda Hoederath, MD
Department of Consultation-Liaison Psychiatry, University Hospital Zurich
Ramistrasse 100, 8091 Zurich, Switzerland
E-mail: linda.hoederath@usz.ch