

BIPOLAR MOOD DISORDER AND MENTAL RETARDATION COMORBID WITH ARNOLD-CHIARI MALFORMATION: CASE REPORT

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ABSTRACT

Arnold Chiari malformation (ACM) is a rare disorder characterized by downward herniation of the brainstem and cerebellar tonsils through the foramen magnum. To date; three cases have been reported with an anxiety disorder and one case has been reported with a schizophrenia like psychotic disorder comorbid with this malformation. In this report we aimed to present a patient who had surgery for ACM with syringomyelia one year before psychiatric consultation and was hospitalized for lack of compliance to treatment and loss of functioning with exacerbation of psychiatric symptoms to a psychiatric ward. She was diagnosed as having bipolar affective disorder, mixed episode. The medication regimen was as follows: quetiapine 600 mg/day, aripiprazole 30 mg/day, lithium 900 mg/day, propranolol 40 mg/day. The patient was discharged after 8 days of remission.

Keywords: Arnold Chiari Malformation, bipolar disorder, mental retardation

ÖZET

Arnold-Chiari Malformasyonu ile Komorbid bir Bipolar Duygudurum Bozukluğu Vak'ası

Arnold Chiari malformasyonu (ACM) beyinsapı ve serebellar tonsillerin foramen magnum boyunca fıtıklaşmasıyla karakterize, nâdir görülen bir bozukluktur. Şimdiye kadar ACM ile eşanlı olarak üç anksiyete bozukluğu vak'ası ve bir şizofreni benzeri psikotik bozukluk vak'ası bildirilmiştir. Bu vak'a takdiminde psikiyatrik başvurudan önceki bir yıl içinde sirengomyeliyle beraber ACM için opere edilen ve yineleyen karma epizodlarla seyreden bir bipolar bozukluk vakası sunulmuştur. Hastanın tedavisi ketiyapin 600 mg/gün, aripiprazol 30 mg/gün, lityum 900 mg/gün ve propranolol 40 mg/gün olarak düzenlenmiş ve 8 günlük remisyondan sonra taburcu edilmiştir.

Anahtar Kelimeler: Arnold Chiari Malformasyonu, bipolar bozukluk, mental retardasyon

INTRODUCTION

Arnold-Chiari malformation (ACM) is characterized by herniation of the cerebellum at various extents through the foramen magnum and may be associated with myelomeningocele and myelodysplasia resulting in various clinical presentations (Paul et al 1983). In the type I malformation there is caudal descent of

the cerebral tonsils only; while in type II the cerebellar vermis and possibly the fourth ventricle and pons are involved. Chiari III represents an encephalocele (external sac containing brainstem and posterior fossa contents); thus, the cerebellum and brainstem are descending not only into the spine, but also into an external sac. Chiari IV consists of cerebellar hypoplasia

(Strayer 2001).

It is more common in females, with a female-to-male ratio 3:1. ACM is usually seen in middle-aged adults (Milhorat et al 1999). The exact cause of the Chiari I malformation is unknown, although there is some evidence for a defect of the paraxial mesoderm resulting in a small, shallow posterior cranial fossa and brainstem and cerebellar herniation through the foramen magnum (Menezes 1995).

Mental retardation with or without epilepsy is reported in ACM I patients (Brill et al 1998). To date; three cases have been reported with an anxiety disorder (Iwabuchi et al 1985, Chisholm et al 1993, Caykoylu et al 2008), and one case with a schizophrenia like psychotic disorder (Ilankovi et al 2006) comorbid with this malformation. Here we reported a patient with bipolar affective disorder, mixed episode with a history of manic and mixed episodes for 15 years comorbid with ACM.

CASE REPORT

A 45 year-old, white woman, married, with three children, uneducated, and unemployed presented to our psychiatric outpatient unit with complaints of anxiety, insomnia, irritability and self-mutilation. She was then hospitalized for serious loss of functioning and self-induced injuries. Although she went to a primary school for 5 years she remained illiterate and got married at 17 years of age. Her family reported that she never had an episode of mood disorder until the age of 30. She was working as a cleaner 15 years ago until she had a mood episode when she got fired where she had worked for ten years. The patient's first psychiatric complaints including restlessness, decreased need for sleep, an increase in spending money, irritability and increased libido emerged fifteen years ago and she was hospitalized and treated with haloperidol. After discharge she stopped taking the medication and was lost to follow up. In 1994, she had another episode with the complaints of decreased need for sleep, irritability similar to her first episode. She was hospitalized again and discharged with remission on haloperidol. Despite her irritability and lack of compliance with treatment, she somehow functioned well between 1994 and 2007. In 2007, she presented to a neurosurgical outpatient clinic with complaints of headache and numbness in her left arm and MRI scans demonstrated ACM-I (tonsillar herniation, 8 mm) and syringomyelia. She underwent an occipital craniotomy, C1 posterior laminectomy surgery along with duraplasty. There was no sign of a neurologic de-

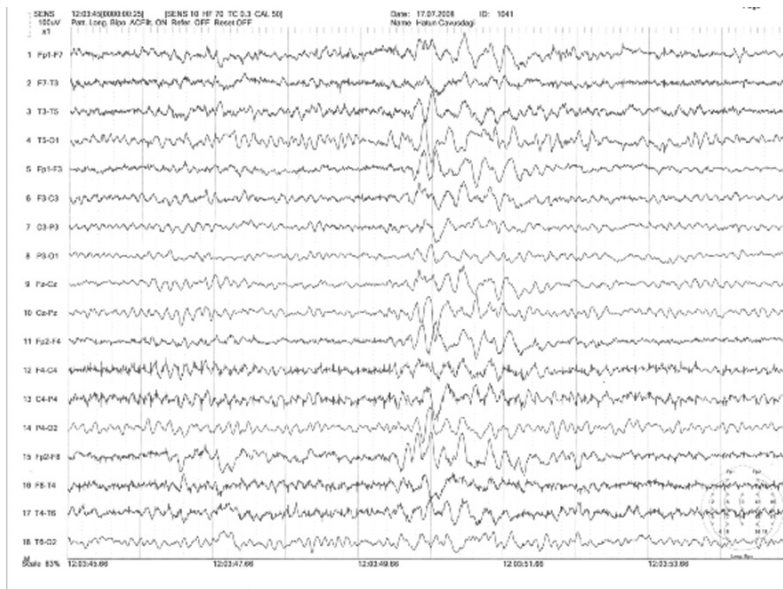
ficit after surgery, but just after the operation she had complaints of hypervigilance, fear, inner discomfort, spasms in both arms and in July 2007 symptoms of anxiety emerged. In February 2008, she was hospitalized in our inpatient clinic with the symptoms of irritability, decreased need for sleep, increased speech, increased libido and hallucinations such as shedding light on her eyes and seeing spiders. She was diagnosed with bipolar disorder type I, manic episode and electroencephalographic (EEG) examination revealed mild (7 to 7.5 Hz) slow background activity and generalized epileptiform discharges provoked by hyperventilation. She was treated with a regimen of valproate 1000 mg/day, quetiapine 100 mg/day and risperidone 3 mg/day, but because of a decrease in white blood cell count, valproate was switched to 1200 mg/day lithium. In July 2008, she presented again with symptoms of irritability, talking to herself, inner discomfort, decreased need in sleep, and loss of functioning and self-harm. Psychiatric examination revealed irritable mood, decreased self care, increase in speech speed, grandiosity, overvalued ideas, negative thoughts about her illness, some worries about her relatives and an increased libido. She was again diagnosed with bipolar disorder type I, mixed episode. IQ test (Porteus) score was 68. Risperidone treatment was switched to aripiprazole 20 mg/day and lithium 900 mg/day due to an increase in prolactin level. The patient was discharged after 8 days with remission.

DISCUSSION

To date; three cases have been reported with an anxiety disorder (Iwabuchi et al 1985, Chisholm et al 1993, Caykoylu et al 2008), and one case has been reported with a schizophrenia like psychotic disorder (Ilankovi? et al 2006) comorbid with ACM.

Our case was diagnosed with bipolar disorder type I, mixed episode with a history of manic and mixed episodes for 15 years.

Psychiatric symptoms may be caused by the ACM which may lead to compression of the brainstem. Serotonergic systems in the dorsal and median raphe nucleus may be part of a distributed neuronal system which regulates anxiety (Spiga et al 2006). Serotonin (5-HT) modulates various neuronal activities, and consequently regulates several physiological and behavioral functions such as the control of impulses, aggressiveness and suicidality (Shiah and Yatham 2000). Decreased 5-HT activity may be associated with a number of abnormalities, such as suicidal attempts, aggressiveness and sleep disorders, all of which are



frequently seen in bipolar disorder (Ackenheil 2001).

Although it has been reported that brainstem lesions in ACM can lead to changes in neurotransmitter levels, other psychiatric disorders are not commonly seen with ACM; so this case should be considered as a coincidence of ACM and bipolar disorder as this is the first case in literature.



Patients tend to do poorly after corrective surgery for ACM if their symptoms persist more than 24 months (Dyste et al 1989). Thus in our case bipolar disorder was not resolved.

Bipolar disorder should be considered when anxiety and uncontrolled impulsions are seen with ACM. After surgical decompression, anxiety and symptoms of mood disorder do not disappear, but remission can be seen with psychotropic treatment. Further clinical studies are needed to illuminate the underlying neuropathogenic mechanisms that contribute to psychiatric disorders in ACM.

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