

Case report / Olgu sunumu**Coincidental mega cisterna magna with psychotic disorder:
a possible neuroanatomical liability for a shared psychotic disorder****Yasin Hasan BALCIOĞLU,¹ Simge Seren KIRLIOĞLU,¹
Tonguç Demir BERKOL,¹ Güliz ÖZGEN²****ABSTRACT**

Mega cisterna magna (MCM) is a cystic malformation of the posterior fossa and characterized by enlarged cisterna magna with an intact vermis and cerebellar hemispheres. The lesion is frequently seen as isolated however it may also constitute a part of Dandy-Walker complex. The concurrence of MCM and psychiatric disorders has already been documented but the possible pathophysiological associations still remain elusive. We present a 36 years-old male patient with incidentally detected isolated MCM, who previously had persecutory and reference delusions as a possible non-index case of shared psychotic disorder. Shared psychotic disorder (folie a deux) is a relatively rare mental condition develops between two affected individuals in a close relationship. In this entity, the index patient already has a psychotic disorder and transmits delusional beliefs on a healthy -secondary- individual. It is firmly suggested that abnormalities of cerebellar structures could involve the pathophysiology of psychotic disorders and based on this fact, we aimed to contribute to the regarding literature with discussing the predisposing role of cerebellar neurodevelopmental abnormalities in psychosis with presenting a possible shared psychosis case. (*Anatolian Journal of Psychiatry* 2018; 19(1):106-109)

Keywords: cerebellum, mega cisterna magna, psychotic disorders, shared psychosis

**Mega cisterna magna ve psikotik bozukluğun rastlantısal birlikteliği:
Paylaşılmış psikotik bozukluk için olası bir nöroanatomik yatkınlık****ÖZ**

Mega sisterna magna (MCM), sisterna magnada genişleme ile birlikte yapısal olarak sağlam vermis ve serebellar hemisferler ile karakterize, posterior fossanın kistik bir malformasyonudur. MCM, izole olarak görülebileceği gibi aynı zamanda Dandy-Walker kompleksinin bir parçası olarak da saptanabilir. MCM ve psikiyatrik hastalıkların birlikteliği bildirilmiş olmasına rağmen, bu tanımlar arasındaki olası patofizyolojik ilişki henüz yeterince vurgulanmamıştır. Bu yazıda, rastlantısal olarak izole MCM saptanan, geçmiş kötülük görme ve alınma sanrıları bulunan ve olası bir paylaşılmış psikotik bozukluğun ikincil olgusu durumundaki 36 yaşında bir erkek hasta bildirilmiştir. Folie a deux veya paylaşılmış psikotik bozukluk, yakın ilişki içinde bulunan iki bireyin etkilendiği görece nadir ruhsal bir durumdur. Bu klinik durumda, halen psikotik bir bozukluğu olan birincil hasta kendi sanrılarını sağlıklı -ikincil- bireye aktarır. Serebellar yapı anormalliklerinin psikotik bozuklukların patofizyolojisinde rol oynadığının öne sürülmesinden hareketle, olası bir paylaşılmış psikotik bozukluk olgusu sunularak serebellar nörogelişimsel anomalilerin psikoza yatkınlık oluşturan rolleri tartışılmış ve literatüre katkı sağlamak amaçlanmıştır. (*Anadolu Psikiyatri Derg* 2018; 19(1):106-109)

Anahtar sözcükler: Mega cisterna magna, paylaşılmış psikoz, psikotik bozukluklar, serebellum

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INTRODUCTION

Mega cisterna magna (MCM) is a developmental variation of the posterior fossa which includes enlarged cisterna magna, morphologically intact vermis and cerebellar hemispheres and occasionally becomes a part of several syndromes such as Dandy-Walker (DW) spectrum disorders.¹ Although this entity had previously been demonstrated in co-occurrence with a number of neuropsychiatric disorders,² remarkable explanation has not been introduced yet. The cerebellum is connected to many regions of the cerebral cortex by the cortico-cerebellar-thalamic-cortical circuit (CCTCC) and known as a crucial actor in this circuit, particularly in equilibration and integration of motor functions. To date, its contribution to the modulation of higher order functions has been increasingly recognized. Novel findings pointed the involvement of the cerebellum and posterior fossa lesions in the pathophysiology of psychotic disorders. The main argument is that diminished Purkinje cell size and reduced excitatory input lead cerebellar and CCTCC dysfunctions those responsible for schizophrenia symptomatology, supporting the conventional assumption of an association between congenital or acquired cerebellar lesions and cognitive/behavioral disturbances.^{2,3} Shared psychotic disorder, known as folie à deux (the folly of two), is an uncommon psychotic disorder in which a healthy person (secondary/non-index case) presents similar disordered ideas of a person with a psychotic disorder (primary/index case).⁴ The credibility of this diagnosis is frequently interrogated by the professionals and diagnostic manuals. This lack of clarity is undoubtedly supported by objective limitations in the etiological and the epidemiological understanding of this entity, as well as the limited knowledge of its natural prognosis; nevertheless, kinship, social isolation, passive personality, cognitive impairment, language difficulties and stressful life events are recognized as predisposing factors.⁴ To the best of our knowledge, there is a much-scanted data describing the association of MCM with psychotic disorders; the arguments seemed to be limited to single case reports, indeed. We reported a coincidental MCM in a psychotic disorder case, meanwhile as a non-index case of a possible shared psychotic disorder, intriguingly.

CASE

We present a 36 years-old man, who was single, high school-graduated and working as an ac-

countant. He was hospitalized through compulsory treatment order due to an offense of reckless injury that he committed two years ago. At that time, he was working as an entrepreneur in a high-yielded investment business with his younger brother. Within that period, both had developed skeptical, persecutory and referential beliefs over their investors. They had thoughts of being insidiously watched and pursued. Two months after the onset of his delusions, in the wake of his intensive, distractive and stressful thoughts, he intended self-mutilative cuts with a knife; however, his mother accidentally got wounded whilst her attempt for prevention. Public prosecution initiated consequently, and his criminal liability was removed with a diagnosis of a psychotic disorder. A verdict of compulsory psychiatric treatment was given. The patient's first psychiatric admission was two years ago because of aforementioned self-mutilation attempt. He received electroconvulsive therapy with antipsychotic medication for three weeks with a diagnosis of psychotic disorder with persistent self-harm ideas. Soon after the treatment, he quit his then business and his paranoid thoughts apparently started to improve. In his current psychiatric examination in our institution, he appeared his age and self-care was normal. Cooperation and orientation were intact. His affect was anxious and the mood was euthymic. Speech output and speed were normal. Any delusion or hallucination was not presenting; however, his strict encapsulated beliefs regarding past persecutory thoughts about his past business colleagues were revealed with a detailed psychiatric interview. Clinic Global Impression Scale score was 7, The Mini-Mental State Examination score was 29, Positive and Negative Syndrome Scale Score was 8. Rorschach's inkblot test did not indicate any psychotic thought content or process. In routine scanning, cranial magnetic resonance imaging (MRI) showed mega cisterna magna in the posterior fossa. The cerebellar vermis and hemispheres were intact with the normal volumes. The remaining structures of the posterior fossa and cerebral cortex were radiologically normal (Image 1). Blood screening and electroencephalogram were carried out and nothing abnormal was detected. Neurological examination was normal, cranial nerves were intact. Neurology referral did not demonstrate any pathology. He had no other physical complaints. His younger brother and mother were interviewed three times. His brother was 32 years old and had no psychiatric history; however, his paranoid delusions regarding their

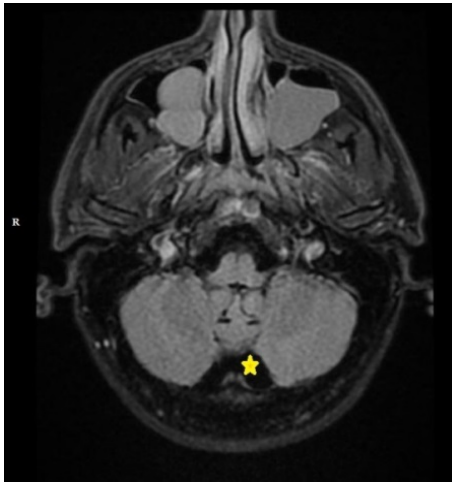


Image 1. Enlarged cisterna magna in MRI
Axial T2 FLAIR

past business colleagues were still presenting. He even asserted being pursued by the same people at present. Their mother claimed that she had not realized any unusual or suspicious incident regarding her sons' allegements and she thought there was no reasonable explanation for business colleagues to pursue them. Social study report did not reveal any unusual and suspicious relationship or incident between the patient and his colleagues, friends, and relatives. The antipsychotic maintenance treatment was started as risperidone 3 mg/day per oral and quetiapine 300 mg/day per oral with the diagnoses of psychotic disorder, not otherwise specified and possible shared psychotic disorder. We thought that his brother was an undiagnosed psychotic disorder case so we suggested to the patient to separate his house and business from his brother. He tolerated the medication well and discharged from the hospital with the current antipsychotic treatment. Informed consent was obtained from the patient in order to present his medical records and psychiatric findings. Routine psychiatric follow-ups were offered after discharge.

DISCUSSION

Abnormalities in cerebellar structures, particularly in vermis, have been implicated to be related to the pathophysiology of psychiatric disorders such as schizophrenia.⁵ The disordered connections mediated through the thalamus between cortical regions and the cerebellum, which are the essential pathways in CCTCC, has been

suggested to explain psychotic symptomatology.¹ Neuroimaging studies revealed a correlation between cerebellar grey matter volume reduction and hallucinations, also between weakened cerebellar activation and delusions in schizophrenia.⁶ The current report is one of the isolated MCM cases presenting with neuropsychiatric manifestations. The epidemiologic and prognostic significance of isolated MCM has not well defined yet; however, normal cognitive functioning is expected. Developmental delay, somatization, and personality disorders were reported with MCM.² The accurate incidence of isolated MCM has not been reported in the general population due to its asymptomatic nature. Enlargement of the cisterna magna is usually associated with cerebellar hypoplasia and ventriculomegaly, therefore, it is critical to exclude ventriculomegaly, cerebellar hypoplasia, posterior fossa lesions or else prior to diagnosing.⁷ Our patient had not suffered from any neurological complaint. His scanning indicated an isolated MCM without any other cranial structural abnormality. MCM and other posterior fossa lesions reported with various cognitive deficits in several cases in the literature;⁷ nevertheless, the patient's cardinal cognitive functions, such as orientation, temporal and working memory, abstracting and computing were well-preserved in the mental status examination. His strict but encapsulated delusions may support the finding that positive symptoms, especially delusions, are frequent in schizophrenia patients with cerebellar lesions.⁸ The shared psychotic disorder is an entity being questioned as for its diagnostic validity. As well as the diagnostic reliability, etiology of shared psychotic disorder still needs to be clarified; heredity, biological, psychodynamic and environmental factors were asserted to be involved.⁴ Unlike the etiology, the first-line therapeutic option of a folie à deux is well-known; separation, combined antipsychotic medication augments the treatment.⁴ Once separation of the business with his brother, our patient's persecutory delusion started to remit. To date, convergent evidence has been provided on underlying cerebellar dysfunction in psychosis. The concurrence of psychosis and MCM in our patient may be attributed to a neurodevelopmental abnormality in the posterior fossa. Although our patient's delusions have been encapsulated, it may also be speculated that structural posterior fossa lesions such as MCM lead dysregulated corticocerebellar neuronal pathways, consequently coming up with a liability of psychosis.

In addition, as a non-index side of a possible shared psychotic disorder, acquiring delusions by a transmission from a psychotic patient may arise as a consequence of this liability. Nevertheless, it would be rather difficult to directly link the occurrence of a shared psychotic disorder to a neuroanatomical abnormality. This article ultimately aimed to demonstrate and discuss a possible predisposition to psychotic symptoms in

the presence of a neurodevelopmental posterior fossa lesion. The incidence of the folié a deux was hypothetically attributed to this predisposition, though. As more studies are reported, a substantive association between psychiatric disorders and MCM, additionally with their hereditary aspects, would find an opportunity to be argued in the future.

Authors's contributions: Y.H.B.: case follow-up, literature review, manuscript writing; S.S.K.: literature review, manuscript writing; T.D.B.: critical review; G.Ö.: case follow-up, critical review.

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