

# Schizencephaly associated with bipolar II disorder

Arora M, Praharaj S K, Sinha V K, Sarkar S

## ABSTRACT

**A 55-year-old man with congenital hemiparesis of the right side, three episodes of generalised tonic-clonic seizure at 16 years of age, and two episodes of severe depression and two episodes of hypomania in the past, presented with severe depression with psychotic symptoms. Computed tomography of the brain showed a grey matter-lined cerebrospinal fluid-filled cleft in the left cerebral hemisphere, involving the temporoparietal region. He was diagnosed to have bipolar II disorder, and was currently severely depressed with psychotic symptoms and schizencephaly. He improved with sodium valproate 1,000 mg/day, quetiapine 450 mg/day and escitalopram 20 mg/day after three weeks without any emergent side effects, and was maintaining well at three months follow-up. Although uncommon, schizencephaly may be considered as one of the differentials in cases of bipolar disorder along with congenital hemiparesis, mental retardation and/or seizures; and neuroimaging should be done to confirm the diagnosis.**

**Keywords: bipolar II disorder, depression, hemiparesis, schizencephaly, seizure**

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## INTRODUCTION

Schizencephaly is a neurodevelopmental disorder originally described by Yakovlev and Wadsworth,<sup>(1,2)</sup> characterised by a cleft in the cerebral mantle, extending from the subarachnoid space to the ventricular system. Schizencephalic clefts are found mostly around the sylvian region and are of two types: type I or closed-lip clefts, with walls opposed to each other; and type II or open-lip clefts, with separated lips and communicating hydrocephalus. They have also been grouped as either unilateral or bilateral clefts.<sup>(3)</sup> Associated malformations include dysplasia of the corpus callosum, absent septum pellucidum, septo-optic dysplasia and arachnoid cysts.<sup>(4)</sup> The clinical features are variable, and include motor deficits (hemiparesis or tetraparesis), seizures (partial or generalised), and mental retardation (mild to severe). The severity mainly depends on the size and location of the clefts, and the presence of associated cerebral

malformations.<sup>(4)</sup> Psychiatric disorders like schizophrenia and bipolar disorder have been rarely reported with schizencephaly.<sup>(5-7)</sup>

## CASE REPORT

A 55-year-old man presented with decreased social interaction, depressed mood, worrying about the future, disturbed sleep and appetite, and denial of the existence of limbs and vital organs for the past two months. He had two episodes of severe depression and two episodes of hypomania in the past. The first episode occurred at the age of 41 years. The hypomanic episodes were characterised by decreased sleep, increased energy, increased confidence and euphoric to irritable moods lasting for a period of 2–3 weeks. He was delivered at term with congenital hemiparesis of the right side, had normal milestones of development and his scholastic performance was average. He had three episodes of generalised tonic-clonic seizure at 16 years of age, but it did not recur thereafter without any treatment. At 18 years of age, he became dependent on alcohol, which changed to occasional intake after ten years. There was no history of bipolar disorder or psychosis in the family.

On physical examination, he had right-sided hemiparesis with circumduction gait, grade 3 muscle power, and brisk tendon jerks on the right side. Mental status revealed an agitated patient, with depressed affect, persecutory and nihilistic delusions. His haemogram, blood glucose, electrolytes, liver and renal function tests, and electrocardiogram were normal. Computed tomography of the brain showed a grey matter-lined cerebrospinal fluid-filled cleft in the left cerebral hemisphere, involving the temporoparietal region. He was diagnosed as a case of bipolar II disorder, currently severely depressed with psychotic symptoms, and schizencephaly. He was admitted to the hospital and treated with sodium valproate 1,000 mg/day, quetiapine 450 mg/day and escitalopram 20 mg/day. He showed a rapid and good response to the treatment after three weeks with resolution of the psychotic symptoms, improvement in mood and psychomotor activity without any emergent side effects. He was maintaining well during follow-up at three months.

## DISCUSSION

In patients with unilateral clefts, hemiparesis of the

Psychiatric Diseases  
Hospital,  
Government Medical  
College, Jammu,  
Jammu & Kashmir,  
India

Arora M, MD, DPM  
Lecturer in Psychiatry

Central Institute of  
Psychiatry,  
Kanke,  
Ranchi,  
Jharkhand 834006,  
India

Praharaj SK, MD, DPM  
Senior Resident

Sinha VK, MD, DPM  
Professor

Sarkar S, MD, DPM  
Senior Resident

**Correspondence to:**  
Dr Manu Arora  
Tel : (91) 191 246 0348  
Fax: (91) 191 258 4226  
Email: drmanu2004@  
rediffmail.com

contralateral hemisphere is seen, and tetraparesis has been reported in bilateral forms.<sup>(4)</sup> Mental retardation is common with bilateral clefts, whereas intelligence is found to be normal in two-thirds of cases with unilateral schizencephaly. The severity of cognitive deficits also depends on the size and position of the cleft; the larger the cleft, the more severe is the deficit. Therefore, patients with closed-lip schizencephalies may not be diagnosed until adulthood, as these individuals have normal intelligence and mild contralateral hemiparesis, as seen in our case.

There have been three previous reports of schizencephaly associated with psychosis,<sup>(5)</sup> two cases of schizoaffective disorder, depressive type,<sup>(6)</sup> one case of bipolar psychotic depression,<sup>(7)</sup> and one case of bipolar I mania with psychotic symptoms.<sup>(8)</sup> To the best of our knowledge, this is the first case in which schizencephaly is associated with bipolar II disorder; the patient had two episodes of psychotic depression in the past and two episodes of hypomania, which responded to treatment. Out of three previous cases presenting with psychotic depression, two had unilateral schizencephaly on the left side similar to our case, while one had bilateral clefts.

Structures that have been implicated in the pathophysiology of bipolar disorder include the temporal cortex, among others.<sup>(9)</sup> Malformations in the entorhinal cortex have been observed in patients with bipolar disorder and major depression,<sup>(10)</sup> lending further evidence to the involvement of the temporal lobe. Furthermore, temporo-occipital cortical dysplasia has been reported to be associated with rapid-cycling bipolar disorder and learning disability.<sup>(11)</sup> In bipolar II disorder patients, abnormalities in the right superior and middle temporal gyri, cingulate gyrus, precuneus and adjacent frontal and parietal white matter abnormalities were reported in high resolution magnetic resonance imaging studies, which correlated with the change in IQ.<sup>(12)</sup> These studies suggest that both bipolar and major depressive mood disorder might be associated with circumscribed neurodevelopmental disturbances in temporal regions. Schizencephaly can be considered to be a severe form of cortical dysplasia, in which the cleft extends all the way into the lateral ventricle.<sup>(13)</sup> In our case, it involved the temporoparietal cortex, which can be hypothesised to

contribute to bipolarity as extrapolated from the above findings.

Although uncommon, schizencephaly might be considered as one of the differentials in cases of bipolar disorder along with hemiparesis, mental retardation and/or seizures. Neuroimaging, preferably magnetic resonance imaging, should be done to confirm the diagnosis. Functional neuroimaging studies in such cases might shed further light on the pathophysiology of bipolar disorder.

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