COMMENT & CRITIQUE

Adult asymptomatic case of Dandy–Walker syndrome associated with bipolar disorder

To the Editor:

Dandy–Walker syndrome is a rare congenital brain developmental malformation (1) that on structural imaging is typically characterised by hypoplasia of the cerebellar vermis and dilatation of the fourth ventricle with a posterior fossa cyst. It invariably results in hydrocephalus and clinically, symptoms are usually observed during the prenatal period or in early infancy. Dandy–Walker syndrome can also occur in association with other brain or systemic malformations, such as corpus callosum agenesis, aqueductal stenosis, syringomyelia, neurocutaneous melanosis and malformations of the heart, face, kidney, limbs, fingers and toes. Recently, Papazisis et al. (2) reported a case of a young man with early onset schizophrenia comorbid with obsessive–compulsive disorder and a Dandy–Walker variant. However, to date, Dandy–Walker syndrome has not been described in association with bipolar disorder, and therefore we briefly report a case of asymptomatic of Dandy–Walker syndrome in the context of bipolar II disorder.

A 40-year-old man, admitted to hospital because of a severe depressive episode, developed after 1 month of antidepressant treatment with venlafaxine, hyperactive and elated mood that earn him the diagnosis of bipolar II disorder and led to the prescription of a mood stabiliser.

On admission, his mental status examination revealed psychomotor retardation and depressed mood, but no delusional ideas or hallucinations. His manner was friendly and cooperative and he remained insightful. His main clinical complaints were anhedonia, insomnia and anorexia. A complete physical and neurological examination was unremarkable. Exploration of the patient's developmental history did not reveal anything unusual and there was no history of familial psychiatric illness.

After providing informed consent, he agreed to blood tests that assessed liver, renal and glandula thyreoidea function and gonadal hormone levels; an electrocardiogram; an echocardiograph; an abdominal ultrasound and a 3D magnetic resonance imaging (MRI) scan. He was prescribed lithium, seroquel and sertraline and after 2-week treatment underwent neuropsychological assessment that included the Wechsler Adult Intelligence Scale-Chinese Revised and Wechsler Memory Scale Revised in China (WMS-RC).

After careful examination, no significant abnormalities were detected except adiposis hepatica, chronic cholecystitis and gallstones. The patient's brain function was within the average range with full-scale IQ scores 98 and 101 scores for WMS-RC.

Interestingly, the MRI scan of the patient showed hypoplasia of the cerebellar vermis, dilatation of the fourth ventricle and enlargement of the rest of the ventricular system with hydrocephalus (Fig. 1A–C). These findings showed that the patient had Dandy–Walker syndrome.

To our knowledge, this is the first recorded case of bipolar disorder in an adult man with Dandy–Walker syndrome. Clinically it is characterised by mental retardation, cerebellar ataxia and symptoms of hydrocephalus. Symptoms, which often occur in early infancy, include slow motor development and rapid increase in head circumference with abnormal bulging at the back of the skull. In older children, symptoms of increased intracranial pressure such as vomiting, irritability, convulsions and signs of cerebellar dysfunction such as unsteadiness, lack of muscle coordination or jerky movements of the eyes may occur. There may also be problems with the nerves that control the eyes, face and neck as well as abnormal breathing patterns. This patient is considered to have adult asymptomatic Dandy–Walker syndrome, as none of the aforementioned symptoms were experienced.

Adult asymptomatic presentation of Dandy–Walker syndrome is extremely rare. Investigation suggested that preservation of the cortical cytoarchitecture as well as the paucity of additional neurodevelopmental changes may explain the absence of clinical expression (3). Judging from the patients' symptoms, the comorbidity of Dandy–Walker syndrome and bipolar disorder is likely to be coincidental. However, evidence for the involvement of cerebellar abnormalities in the pathophysiology of bipolar disorder may suggest a direct causal relationship. The cerebellum plays an important role in cognition and psychiatric disorders (4) and, several morphometry neuroimaging studies have identified significantly smaller vermal subregion volume in bipolar disorder subjects, especially those that have experienced multiple episodes (5–6). Moreover, Lauterbach (7) reported that cerebellar lesions may induce mania and diminished cerebellar outputs may result in abnormal neuronal oscillation in bipolar disorders, especially in rapid-cycling bipolar disorder. In addition, some brain metabolites such as *N*-acetylaspartate (NAA) and creatine (Cr) have been found to be approximately 8% lower for children with a mood disorder than healthy children within the cerebellar vermis (8). In sum, cerebellar abnormalities may be a risk factor for bipolar disorder, while recurrent episodes perhaps aggravate cerebellar atrophy. Clearly, further examination of this brain region is needed along with heightened awareness of this potential significance in the pathophysiology of mood.

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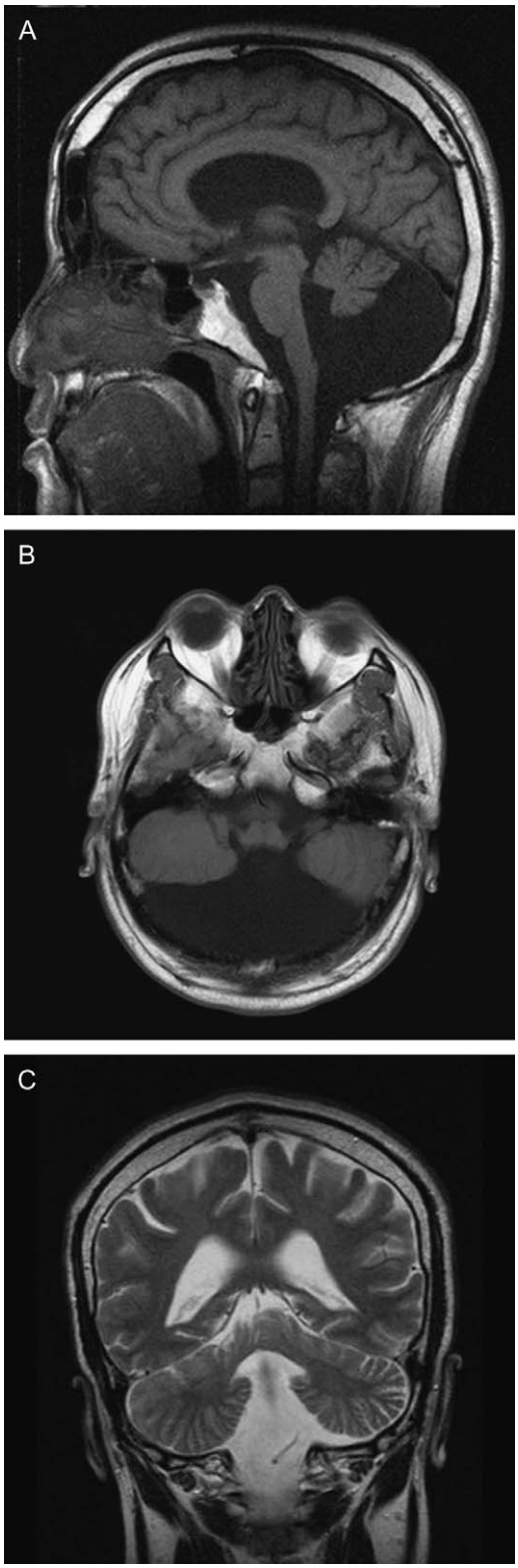

Fig. 1. Magnetic resonance imaging (MRI) of the patient. (A) Midsagittal view of brain, note severe hypoplastic cerebellar vermis and dilatation of the fourth ventricle with a posterior fossa cyst. (B) Axial T1 MRI view of brain showing hypoplastic cerebellar vermis and dilatation of the fourth ventricle. (C) Coronal T2 MRI view of the same patient, show hypoplastic cerebellar vermis, dilatation of the fourth ventricle and enlargement of the rest of the ventricular system with hydrocephalus.

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