

Psychiatric Manifestations Associated With Mega Cisterna Magna

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The Dandy-Walker variant is a milder form of the Dandy-Walker complex and is characterized by normal-sized posterior fossa, mild vermian hypoplasia, and a cystic lesion that communicates with the fourth ventricle. This syndrome has been described in association with schizophrenia, obsessive-compulsive disorder, manic episode, psychosis (delusional type), and recurrent catatonia. The authors present two cases of mega cisterna magna associated with mania and catatonic schizophrenia.

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Dandy-Walker malformation is a rare congenital abnormality characterized by large posterior fossa cyst with open communication between the fourth ventricle, absent or severely atretic inferior vermis, and enlargement of the posterior fossa with elevation of the confluence of sinuses, lateral sinuses and tentorium.¹ The Dandy-Walker variant (DWV) is a term that is frequently used in clinical practice to describe a milder form of the Dandy-Walker complex that refers to a constellation of findings including a normal sized posterior fossa, mild vermian hypoplasia, and a cystic lesion that communicates with the fourth ventricle.²

There is very limited data that describes the association of mega cisterna magna with mania and schizophrenia. The evidence is limited to single case reports. This syndrome has been described in association with schizophrenia and obsessive-compulsive disorder (OCD),³ manic episode,⁴ psychosis (delusional type),⁵

and recurrent catatonia.⁶ There are no reports describing the association of catatonic schizophrenia with mega cisterna magna. We present here two cases of mega cisterna magna associated with mania and catatonic schizophrenia. Per our knowledge, this is the first report of mega cisterna magna associated with catatonic schizophrenia.

CASE REPORTS

Case 1:

A 26-year-old unmarried man from rural background presented with the chief complaints of logorrhea, inappropriate familiarity, delusion of grandiosity, reduced need for sleep, increased activity, and increased consumption of tobacco and alcohol. He was also found to be abusing family members with minor provocations and started spending more money. All symptoms were acute in onset and gradually progressive for 2 months. The patient was not treated for his illness when he presented to us in the outpatient department. There was no history of hearing of voices, suspiciousness about people, headache, blurring of vision, or vomiting. There was no history of mania, hypomania, or any other psychiatric illness in the past or in the family.

Physical examination of the face revealed low set ears and elongated face. The examination of the nervous system revealed hypotonia, mild in-coordination noted on finger-nose test, no nystagmus, normal power, sensations, and gait. Examination of the eyes revealed normal cornea, retina, and visual acuity. Mental status examination revealed increased psychomotor activity, logorrhea, reduced reaction time, flight of ideas, impaired judgment, and absent insight.

The patient was diagnosed as mania with psychotic symptoms based on the ICD 10 criteria and rated on Young Mania rating scale with the score of 37/60 at admission. The patient was started on lithium 800 mg/day

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and risperidone 4 mg/day. The fourth day after starting the medications, the patient became delirious and had urinary incontinence on the fourth day. The patient was shifted to the intensive care unit, and a physician opinion was sought. Blood investigations revealed serum lithium of 0.9 meq/liter, SGPT 233U/liter and creatine phosphokinase was 569 U/liter. CT scan of the brain showed giant cisterna magna measuring 18×22 mm in the occipital region posterior to cerebellum, communicating with the fourth ventricle through a slit like opening. The vermis and the cerebellar hemispheres were normal. There was no dilatation of the posterior cranial fossa. The patient improved in the next 48 hours with conservative management.

Case 2:

A 20-year-old male student from rural background presented with the chief complaint of poor academic performance, wandering behavior, not taking part in daily activities, and reduced self-care, which was insidious in onset and gradually progressive for about 2 years. Six months ago, he had wandered away without informing the family and was found sleeping on the platform of a bus station; 3 months later he was spotted by the local police who found him sleeping in the same place for about 4 days. When he was brought back by the family members, he was poorly kempt with dirty clothes, bad oral hygiene, and long hair. The details of the different events during that period could not be obtained. On asking repeatedly, occasionally he spoke in one or two words in a whispering voice that was difficult to understand and rapport could not be established with him. There was no history of suspiciousness or hearing of voices, no sadness, disinterest, suicidal ideations, and no history of headache, blurring of vision, vomiting, or substance abuse. There were no similar complaints in the past; he was premorbidly well adjusted. There was no history of psychosis in the family.

Physical examination of the face and the limbs revealed no abnormality. There was no thyroid swelling. The patient was ill kempt and had poor oral hygiene. The detailed neurological examination after the patient improved revealed hypotonia, normal power, no nystagmus, normal sensations, and coordination. On mental status examination, the patient had reduced psychomotor activity. When he was made to walk, he would stand in one place with his head facing downward and would sit in the same place for 1–2 hours. He would slowly bring back his hands to a comfortable resting position when

they were placed in an uncomfortable posture. He also exhibited ambivalence when he was asked to protrude his tongue. When asked repeatedly, occasionally he spoke in one or two words in a whispering voice that was difficult to understand suggesting negativism and mutism. He exhibited blunt affect with no reactivity of mood and retardation of thoughts. A diagnosis of catatonic schizophrenia was made based on the ICD 10 criteria. Bush Francis catatonia rating scale was administered to determine the severity of catatonia, which gave a score of 23/69 at admission.

The patient was started on risperidone 3 mg/day. Within the next 6 hours, the patient developed acute dystonia with twisting of the limbs and extension of the neck. His blood and urine examination did not reveal any abnormality. The patient's EEG was normal; MRI of the brain revealed giant cisterna magna measuring 15×20 mm in the occipital region posterior to cerebellum, communicating with the fourth ventricle through a slit-like opening. Incidentally, the vermis and the cerebellar hemispheres were normal. There was no dilatation of the posterior cranial fossa.

DISCUSSION

Andreasen et al⁷ in 2008 suggested that the cerebellum is connected to many regions of the cerebral cortex by a cortico-cerebellar-thalamic-cortical circuit (CCTCC) and cerebellum may play a crucial role in this distributed circuit and coordinate or modulate aspects of cortical activity. Andreasen et al⁸ in 1999 suggested a neo-Bleulerian unitary model and described schizophrenia as a neuro-developmentally derived "misconnection syndrome," involving connections between cortical regions and the cerebellum mediated through the thalamus. Any abnormality in this circuitry leads to misconnections in many aspects of mental activity or cognitive dysmetria. Andreasen et al⁷ in 2008 opined that a decrease in the Purkinje cell size and decreased excitatory input to them from the granule cells have major implications that explain cerebellar and CCTCC dysfunction in schizophrenia and related abnormalities in symptoms and cognition.

Lauterbach⁹ in 2001 suggested that although the study of cerebellum in primary bipolar disorders has not as yet led to clear findings, the study of secondary bipolar disorders after isolated lesions provide some evidence of cerebellar involvement in mood disorders. It is possible

that cerebello-subcortical circuits are involved in mediating mood disturbances.

The various minor physical anomalies associated with Dandy Walker Syndrome include hypotonia, strabismus, myopia, a short neck, microcephaly, brachycephaly, hypertelorism, antimongoloid slant of palpebral fissures, globulus large nose, large mouth with down turned corners, poorly lobulated ears, high arch palate, cleft palate, small hands and feet, clinodactyly, and the brachymesophalangy of the little fingers.¹⁰ In our case series, only the first case had low set ears and elongated face, and there were no minor physical anomalies seen in the second case.

Brain imaging was considered in both the cases in view of acute side effects even with lower dosages of our medications. Thus, it may be advisable to start the antipsychotic drugs at a lower dosage and titrate slowly in patients with mega cistern magna as they may be more vulnerable for drug-induced extra pyramidal side effects.

Current genetic findings are beginning to provide suggestive evidence that there are genetic loci that contribute susceptibility across schizophrenia, bipolar disorder, and schizoaffective disorders.¹¹ Though the exact etiology for mega cisterna magna, mania or schizophrenia

is not clear, a study by Langarica *et al*⁵ in 2005 suggested that both the psychotic disorders and the mega cisterna magna may be the expression of a single underlying neurodevelopment abnormality.

In our case series, co-occurrence of mega cistern magna and psychiatric disorders (mania and catatonic schizophrenia) could be more likely a chance association or an incidental finding. Rarely, both of these conditions may have a common underlying etiology that may indicate a causal relationship. Further studies are required to establish a causal relationship between the mega cistern magna and the psychiatric disorders.

CONCLUSIONS

Our case series suggest that mega cisterna magna may be associated with psychiatric manifestations like mania or catatonic schizophrenia.

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