

Scoliosis in Dandy-Walker syndrome: a case report and review of literature

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Abstract: This submission presents a case of scoliosis in a patient with established Dandy-Walker anomaly of the brain. A retrospective review of the patient's case notes was undertaken and the limited literature on this subject reviewed. The 13-year-old girl presented with a stiff right thoracic scoliosis typical of adolescent idiopathic scoliosis. The scoliotic segment also presented with significant lordosis. She had facial and truncal dysmorphism characteristic of Dandy-Walker complex and her brain images confirmed the diagnosis. She underwent scoliosis surgery by the posterior approach uneventfully. In conclusion scoliosis is hitherto unreported in the Dandy-Walker complex. The results of intervention appear satisfactory.

Keywords: Dandy-Walker syndrome; scoliosis; thoracic lordosis; Dandy-Walker complex

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Introduction

The Dandy-Walker complex is a rare congenital intracranial malformation that comprises a spectrum of abnormalities of the posterior cranial fossa which are classified as follows (1):

- (I) Dandy-Walker malformation (cystic dilatation of the 4th ventricle, complete or partial agenesis of the cerebellar vermis and an enlarged posterior cranial fossa).
- (II) Dandy-Walker variant (cystic posterior cranial fossa mass with variable hypoplasia of the cerebellar vermis and no enlargement of the posterior fossa).
- (III) Mega Cisterna Magna (enlarged cisterna magna with normal cerebellar vermis and fourth ventricle).

Clinical features

Typically, the patient presents with psychomotor and growth retardation, hypotonia, strabismus, myopia, short neck, microcephaly, brachycephaly, hypertelorism, anti-mongoloid slant of palpebral fissures, large globular nose, large mouth with down turned corners, poorly lobulated ears, high arched palate, cleft palate, small hands and feet, clinodactyly, and the brachymesophalangy of the little

fingers. Spinal deformity has not been described as an association of this disease complex (2,3).

Case presentation

MSAAH is a 13 years old girl who was referred to our spine unit from one of the regional hospitals for evaluation of scoliosis. The patient's mother had observed progressive deformity of her back since 12 months of her visit to our center. She had attained menarche a month before admission to our hospital. She was floppy since birth, microcephalic, had facial dysmorphism, and was an epileptic on medications. On examination she was ambulant with a normal gait. She was asthenic in build, with obvious microcephaly with a flattened occiput, long neck and hypotonia of the limbs with no ligament laxity. She had a facial deformity consisting of a large nose and mouth with multiple un-erupted teeth. There were no neurological deficits in her upper or lower limbs and all her reflexes were normal. Her bladder or bowel functions were also normal. She appeared to have a right thoracic curve with a right-sided rib hump and the same shoulder was slightly depressed. The scapulae appeared dysmorphic with beak-like prominences at the inferior poles (*Figure 1A*). Her

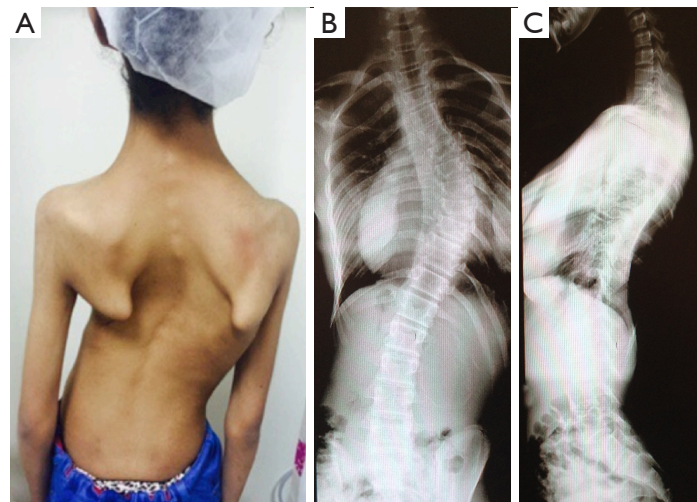


Figure 1 Pre-operative status of the case described. (A) Pre-operative clinical photograph of the patient. Please note the scapular deformity and the thoracic lordosis; (B) pre-operative standing AP X-ray of the whole spine demonstrating the spinal deformity; (C) standing lateral X-ray of the spine. Please note the significant thoracic lordosis. AP, anteroposterior.

standing height was 150 cm, sitting height 74 cm and arm span was 156 cm.

X-rays of the whole spine (standing views) revealed an idiopathic pattern of scoliosis in the thoracic region with apex at D8 and ends at D5 and D12 (she had 13 thoracic vertebrae with ribs and 4 lumbar vertebrae). The main thoracic curve measured 50 degrees Cobb and was correcting to 38 degrees on side bending. The proximal thoracic curve was 17 degrees correcting to 12 degrees and the lumbar curve measured 27 degrees with full correction on bending (*Figure 1B*). The standing lateral view showed the entire thoracic and lumbar spine forming one smooth lordotic curve with the cervicothoracic junction and cervical spine in mild kyphosis (*Figure 1C*).

Her skull CT scan showed an enlarged cisterna magna with normal cerebellum, normal vermis and normal 4th ventricle (*Figure 2A*). The cerebral parenchyma appeared normal with no focal lesions or midline shifts. This suggested a mild form of Dandy-Walker syndrome. She had an MRI of the brain that confirmed these findings (*Figure 2B-D*). She also had a whole spine MRI screen that did not reveal any anomaly. The patient was evaluated by the Neurosurgical team before the surgery and no intervention was recommended.

Surgery

She was planned for posterior scoliosis correction using

the USS II (DePuy-Synthes, Synthes GmbH, Oberdorf, Switzerland) system with an all-screw construct from D4 to D13. At surgery the spine was found to be visibly lordotic in the thoracic region. The rods were over contoured to correct this lordosis but they tended to straighten out (6 mm TAN rods). Correction was achieved by translation, rod rotation and direct vertebral rotation (DVR) maneuvers and the montage was tightened. Intra-operative SSEP and MEP monitoring was performed throughout without any adverse events.

After surgery the patient stayed in the hospital for 4 days during which time she was progressively mobilized and her wound remained clean and dry. She was discharged home on the 5th day and reviewed at 2 weeks for surgical site inspection. Her postoperative X-ray revealed good correction of the coronal plane deformity with good sagittal and coronal balance (*Figure 3A-C*). However, the thoracic lordosis seemed to be under-corrected. Her pre-operative thoracic kyphosis was measured as -23 degrees from D2 to D12 and her post-operative kyphosis as +25 degrees. She was reviewed thereafter in the outpatient clinic as per our protocols at 6 weeks, 12 weeks and 6 months. Her cosmetic and functional results were excellent at last visit (*Figure 3A*).

Discussion

This is the first report of idiopathic scoliosis deformity in Dandy-Walker syndrome or its variants. A detailed literature search was instituted to retrieve similar

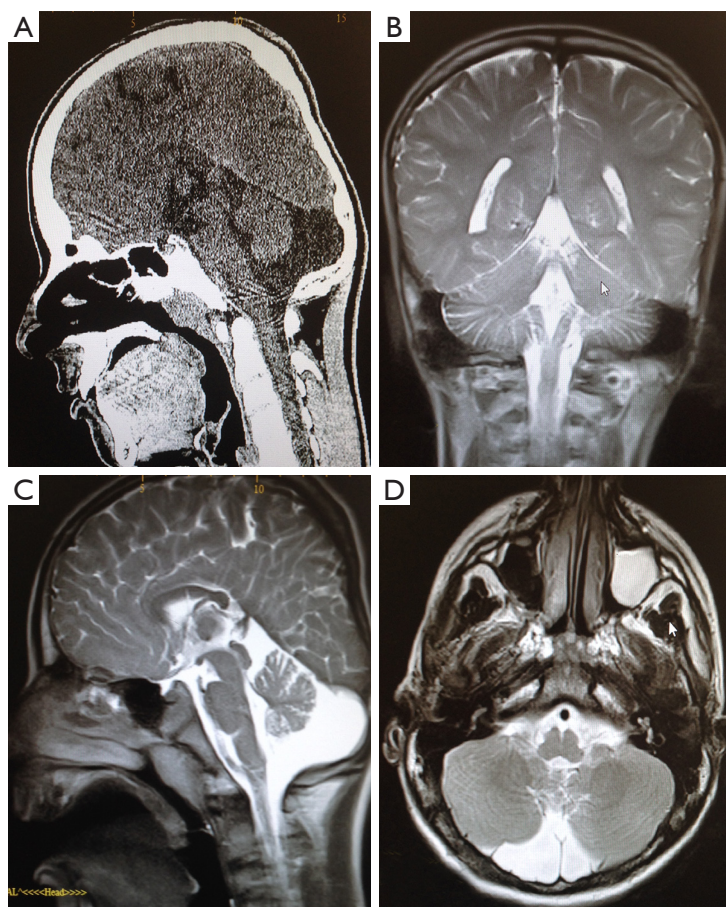


Figure 2 Representative imaging studies of the skull illustrating the Dandy-Walker anomaly. (A) CT scan of the skull illustrates the microcephaly and the flattened occipital region; (B-D) coronal, sagittal and axial MRI scans (T2 weighted images) of the skull clearly demonstrates the enlargement of the cisterna magna.

cases but none had been reported to the best of our knowledge. The Dandy-Walker malformation is a term that represents a spectrum of congenital abnormalities of brain development (1-3). This disorder typically involving the fourth ventricle and the cerebellum was first described in 1914 by W Dandy and AE Walker and was designated as Dandy-Walker syndrome in 1954 by C Benda, who also reported a familial occurrence. The malformation has an estimated prevalence of about 1:30,000 live births with a slight female preponderance and is responsible for 4–12% of infantile hydrocephalus. It is frequently associated with other intracranial anomalies such as agenesis of the corpus callosum, holoprosencephaly, occipital encephaloceles and ocular abnormalities. Extra-cranial anomalies include polycystic kidneys, cardiovascular defects, polydactyly and cleft palate. Postnatal studies indicate that the incidence of

associated malformations range between 50% and 70%.

Scoliosis is a hitherto un-described anomaly associated with Dandy-Walker syndrome though a recent paper reported scoliosis associated with Aicardi syndrome, a congenital absence of the corpus callosum of the brain (4). The clinical appearance of the curve is similar to Idiopathic scoliosis though the curve appears very stiff even at early stages of progression. Idiopathic scoliosis is known to produce lordotic deformity of the affected segments. In the case described the degree of lordosis and the stiffness of the curve in the sagittal plane appeared quite severe in relation to the coronal plane deformity. All other features like vertebral rotation, behavior of the secondary curves, time of onset and trunk symmetry were similar to adolescent idiopathic scoliosis. It is distinctly possible that the spinal deformity was an incidental association and not related to the

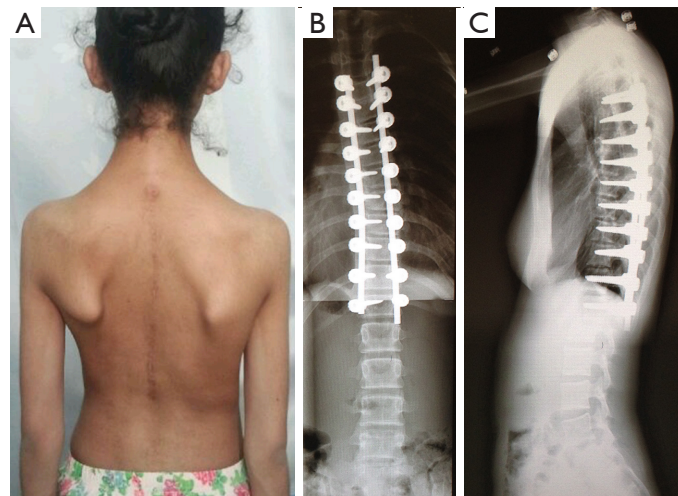


Figure 3 Post-surgery status of the case illustrated. (A) Post-operative clinical photographs of the patient showing excellent cosmetic correction; (B,C) post-operative standing AP and lateral view X-rays of the patient demonstrating the correction obtained and the residual lordosis of the thoracic spine. AP, anteroposterior.

underlying brain disorder. One remarkable feature was the beak-like malformation of the scapulae which has also been unreported thus far; this malformation appears to be exacerbated by the spinal deformity because after correction of the scoliosis the cosmetic appearance of the scapulae seem to have improved substantially (*Figure 3A*).

Conversely, intraspinal anomalies associated with scoliosis have been extensively described in literature (5-9). They are more frequently associated with congenital rather than idiopathic scoliosis and tends to present as Chiari 1 malformations, syringomyelia, tethered cord, low lying conus and tonsillar ectopia. The association of mega cisterna magna in a case of idiopathic scoliosis is also hitherto undescribed, to the best of our knowledge.

Conclusions

Dandy-Walker syndrome and its variants can produce scoliosis which clinically appears similar to adolescent idiopathic scoliosis.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest

to declare.

Informed Consent: Informed consent has been obtained from the subject of this study.

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