

Posterior Kyphectomy in a Myelomeningocele Patient with Gibbous Deformity: A Case Report and Literature Review

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Abstract

Myelomeningocele is a condition caused by the disjunction of neuroectoderm and ectoderm during embryogenesis. It can lead to neuronal structure disorders such as kyphosis and scoliosis, with scoliosis being more common. Kyphosis is found in 10-15% of these patients and can cause poor clinical consequences. The annual progression of Kyphosis is approximately 8 to 12 degrees. Kyphosis is a limiting factor in rehabilitation due to the inability of the patients to sit in wheelchairs and in the apex of the kyphotic region. This study aims to report a similar condition in a 14-year-old Iranian boy who underwent surgery for posterior kyphectomy. In the majority of cases with kyphosis, anterior wedging occurs in the vertebral body where the apex of the deformity is located. In the studied patient, the deformity had a round curve with no definite apex to mark out. These deformities need special surgical approaches and postoperative care.

Key Words: Child, Iran, Myelomeningocele, Kyphosis, Kyphectomy.

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1- INTRODUCTION

Myelomeningocele is a condition caused by the disjunction of neuroectoderm and ectoderm during embryogenesis. It can lead to neuronal structure disorders such as kyphosis and scoliosis, with scoliosis being more common (1). Kyphosis is found in 10-15% of these patients and can cause poor clinical consequences (2). The annual progression of Kyphosis is approximately 8 to 12 degrees (3). Kyphosis is a limiting factor in rehabilitation due to the inability of the patients to sit in wheelchairs and in the apex of the kyphotic region (4). Sharrard first described vertebrectomy in 1968 as a surgical treatment for correcting and improving vertebral alignment in patients with myelomeningocele (5). Nowadays, the classic approach in treating these patients is kyphectomy and posterior instrumentation (6). This study examines a 14-year-old male patient who underwent kyphectomy and fixation due to severe thoracolumbar

kyphosis. The patient had a case of myelomeningocele repaired during his neonatal period.

2- CASE REPORTS

A 14 year-old paraplegic male patient with thoracolumbar Gibbous deformity was referred by an urologist to Akbar Children Hospital in Mashhad, Iran, in August 2020. The patient could not undergo a cystoscopy procedure because he was unable to lie down in the supine position. The patient had bilateral hydronephrosis and renal dysfunction, had needed Clean Intermittent Catheterization (CIC) for many years, and had a history of pyelonephritis as well. In physical examination, he was diagnosed with thoracic deformity, sensory level at about T7, was completely paraplegic under the level of T7, and lower limbs were atrophic. Computed Tomography Scan (CT scan) of abdomen showed that the left kidney was in the kyphotic curve (**Figure.1**).

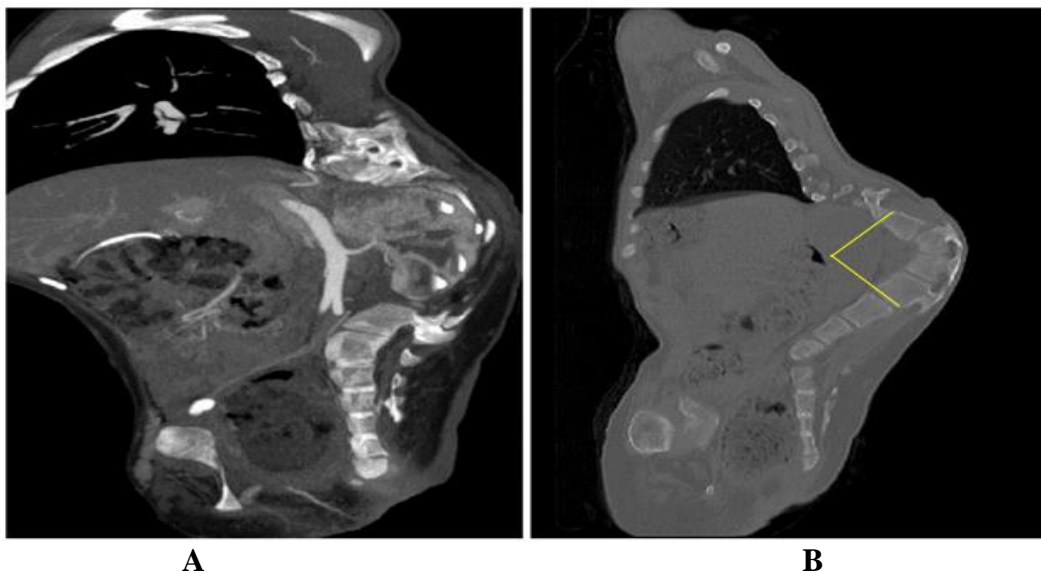


Fig.1: A: Abdominopelvic CT angiography of patient with left kidney in deformity
B: Spinal computed tomography.

2-1. Operation

The surgery was performed with the patient in the prone position, with a roll under the chest and another roll under the hips (**Figure.2**). Previous incision was at the Gibbus deformity region. The level of the previous surgery from T8 to L5 was opened for exposing the site. The skin over the curve had a large midline scar. After opening the fascia and subperiosteal dissection of muscles, pedicle screws were placed in superior vertebrae (T9-T10 –T11), and inferior vertebrae (L3-L4). The Canoe method was used for placing the screws (19). The thecal sac was ligated and then cut at the level of T11. Total spondylectomy of T12, L1 and L2 was performed to expose the Anterior Longitudinal Ligament (ALL).

Then, the spinal reconstruction was performed using rods to place the lower endplate of T11 and the upper endplate of L3 vertebral body tight together. Bone autograft was utilized to fill the gap between the two ends of the spine. The bleeding throughout the operation was estimated at 130 ml and the total duration of the surgery was 5 hours.

2-2. Post-operative condition

Complications such as Cerebrospinal Fluid (CSF) leakage or infections did not occur after the surgery. The patient was able to lie down in the supine position and sit in the wheelchair without difficulty the next day of the operation (**Figure. 3**).



Fig.2: Gibbus deformity.

Note the severe sharp angle thoracolumbar spinal deformity.

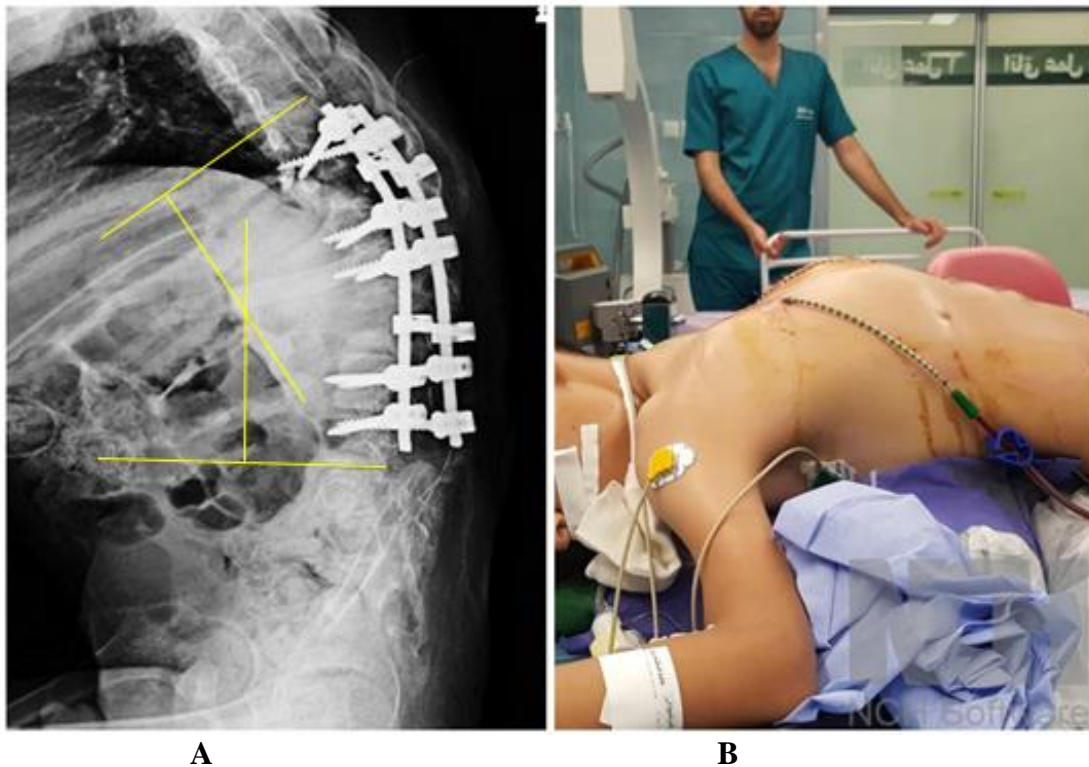


Fig.3: A: Lateral X-ray of the patient, 2 month after surgery.

Note the correction of the deformity.

B: Lateral view of the patient at the end of the surgery.

3- DISCUSSION

Myelomeningocele is a condition caused by the disjunction of neuroectoderm and ectoderm during embryogenesis. It occurs in 0.005-0.2 % of births. Neuronal structure dysfunction can cause kyphosis and scoliosis (1). Spinal deformities are found in 50% of patients (7). Kyphosis is usually seen in thoracolumbar spines and the kyphotic curve is more than 80 degrees in most of the cases (8). Patients are unable to sit upright without the use of their hands because of severe kyphotic deformity. They are also not able to lie down in the supine position due to the Gibbus deformity. Over time, as the deformity progresses, it can lead to a reduction in lumbar vertebral height. These alterations decrease the thoracic compliance due to

the restriction of the thoracic cavity growth and protrusion of the intra-abdominal organs into the thoracic cage. The result of these changes is the compensatory thoracic lordosis to increase the pulmonary function (9). No clinical clue or complaint was found in the patient's pulmonary function at the 6 months follow-up. In addition, our patient was not at risk for thoracic insufficiency syndrome, this complication is seen in younger children who undergo a long instrumentation involving the proximal (involvement of T1 or T2) thoracic spine (20). Treatment of hyperkyphosis is a challenging issue in myelomeningocele patients. This progressive deformity is a result of defects in spinal posterior body elements and lateral vertebral extensors displacement, accompanied with the intensive effect of the spinal flexors that change vertebral

biomechanics and bending forward (10). Skin damages at the Gibbous region, sagittal imbalance, and deformity progression are indications for surgical treatment. Most surgeons recommend the age of 5-12 years for surgery if there is adequate skin at the site of kyphosis (12). The best estimated age for surgery is 8 years and older to prevent the growth restriction due to the long fusion time (9). Kyphectomy surgery is the treatment method of choice in patients with rigid kyphosis and myelomeningocele (13, 14). Sharrard first described vertebrectomy in 1968 as a method for correcting deformities and vertebral alignment (5). Fixing methods for the surgical treatment of kyphosis are: anteroposterior fusion, posterior fusion only, anterior and posterior plating, in situ fusion by Spica cast, Harrington rod, Dwyer instrumentation, luque rod, cable, hook, wire, pin, Gardner bottle screw, Hartshill rectangle, lumbar pedicle screw alone, Isola rod, and Cotrel-Dubousset instrumentation (16).

Kyphosis can be corrected up to 47 degrees after vertebrectomy and placing trans-corporal screws (9). Heydemann and Gillespie were able to correct the kyphosis up to 91.2 degrees compared to the preoperative condition. Huang and Lubicky corrected the kyphotic degree up to 104.5 degrees using modified Gillespie method (17). Ganjeifar et al. (2016) reported a case of severe myelomeningocele-related kyphosis that underwent surgery with an extraperitoneal approach and 5-level en bloc spondylectomy. They had also performed interbody and posterolateral fusion and long segment pedicle screw fixation. According to the authors, this method could reduce the severity of complications such as bleeding and mortality (18). The general complications of surgery include delayed surgical wound healing, bleeding, and occasionally death (10). The incidence

of complications in these surgeries has been reported up to 90% with the most common being infection of the surgical site and wound dehiscence (1). Thin and poorly repaired skin in the kyphotic region can cause recurrent infections and CSF leakage (3). Other complications include significant rates of pseudoarthrosis, and implant failure (15).

4- CONCLUSION

In the majority of cases with kyphosis, anterior wedging occurs in the vertebral body where the apex of the deformity is located. In the patient examined in this study, the deformity had a round curve with no definite apex to mark out. These deformities need special surgical approaches and postoperative care.

5- CONFLICT OF INTEREST: None.

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