



Original Article

Neurosurgical management of Currarino syndrome: A case series and review of literature

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ABSTRACT

Background: The Currarino syndrome (CS), defined by the triad of anorectal malformations, sacral bone deformities, and presacral masses, is rare. There are few surgical series that discuss conservative management versus the surgical approaches to these lesions. Here, we describe utilizing a combined anterior and posterior approach for resecting these lesions in four patients.

Methods: Four patients with CS were treated with two-stage approaches performed by a multidisciplinary team, including pediatric neurosurgery and general surgery. The first anterior laparoscopic approach mobilized the presacral mass from its ventral attachments. The second posterior procedure detethered the spinal cord, repaired the dural defect, and facilitated removal of the presacral mass.

Results: Gross total resection of all four presacral masses was accomplished without intraoperative complication; all patients clinically improved.

Conclusion: The CS is characterized by a large presacral mass. Here, one must rule out malignancy and also consider diagnosis/resection due to the risks for malignant transformation. The operative approach we described in four patients utilized standard anterior mobilization of the mass, followed by posterior detethering, dural repair, and ultimate resection.

Keywords: Currarino syndrome, presacral mass, tethered cord, ventral meningocele

INTRODUCTION

Currarino syndrome (CS) is a rare condition defined by a triad consisting of anorectal malformations, sacral bone deformities, and presacral masses.^[2,7] Approximately 70% of cases are familial with an autosomal dominant inheritance pattern.

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Although over 300 patients with Currarino's triad have been reported/treated, there is no consensus regarding their optimal conservative versus surgical management.^[3] Most surgeons plan their treatment based on the symptoms, physical examination, and radiographic findings. Conservative management may be an option if the patient is essentially asymptomatic despite the presence of sacral anomalies. Others recommend lesion resection due to the presence of symptoms/signs and/or risk of malignant transformation.

Different surgical approaches have been utilized.^[2] Here, we report four sequential patients with symptomatic CS treated successfully with sequential, staged, anterior followed by posterior approaches.

CASE SUMMARIES

Four patients were included in this study [Table 1]. They averaged 12.7 years of age and included two males and two females; the two males were brothers. A family history of anorectal malformation and neural tube defects was identified in three of the four patients. At the time of birth, three of the four patients had anorectal malformations, constipation, and pelvic pain, requiring surgical repair. Two patients (brothers) had a history of imperforate anus and tethered cord. One patient reported pain with forward flexion at the hip, and another reported severe menstrual cramping.

Magnetic resonance studies and surgery

An magnetic resonance (MR) imaging scan was obtained on all patients and revealed presacral masses with sacral defects. All underwent a combined anterior/posterior approach performed by a multidisciplinary team including pediatric neurosurgery and general surgery. The pathology revealed each patient had both a sacral meningocele and teratoma. Only one patient had a perioperative complication consisting of a superficial wound infection treated successfully with antibiotics. Patients were followed an average of 8 postoperative months, by which time all exhibited improvement in their symptoms including the resolution of pelvic pain. Further, constipation resolved completely in one patient and improved but persisted in the other three. None demonstrated delayed neurological deterioration.

SUMMARY OF OPERATIVE TECHNIQUE

Anterior approach

Each patient was initially positioned supine, and laparoscopic access was obtained to the presacral mass. It was carefully dissected away from surrounding structures, with care taken to identify and preserve both ureters. The dissection was taken down to the anal canal and the levators. Following sufficient mobilization of the meningocele, the abdominal wounds were closed, and the patient was turned prone.

Table 1: Summary of demographic and clinical characteristics.

| Case | Sex | Age | Family history | Preoperative deficit | Ultrasound anomaly/MR findings | Surgical procedure | Pathology | Outcome/follow-up |
|------|-----|----------|------------------------|--|---|--------------------------------------|----------------------------------|--|
| 1 | F | 40 weeks | No | Difficulty defecating (required colostomy) and bilateral clubfoot deformities | US: closed neural tube defect S2-S3 MR: large right sacral defect with mass (17 mm×20 mm×27 mm) – fat, fluid, neural tissues extended through sacrum into retroperitoneum | Combined anterior/posterior approach | Teratoma and ventral meningocele | Gross total resection Follow-up at 6 months: normal voiding, no neurologic deficit |
| 2 | F | 16 years | Yes | Hip and leg pain, severe menstrual pain and constipation | MR: mass centered on the S3–S4 left neural foramen (2.4 cm×4.8 cm×4.2 cm), additional presacral mass (10.8 cm×10.7 cm×8.7 cm) – both masses exerted significant mass effect on the uterus, ovaries, and bladder | Combined anterior/posterior approach | Teratoma and ventral meningocele | Gross total resection, superficial wound infection (resolved with antibiotics) Follow-up at 1 year: pain resolved, constipation improving |
| 3 | M | 21 years | Yes, brother of Case 4 | Pain with forward flexion and constipation (history of tethered cord and imperforate anus) | MR: multilocular presacral and precoccygeal mass (5.3cm×2.8×4.3 cm) communicating with sacral subarachnoid space | Combined anterior/posterior approach | Teratoma and ventral meningocele | Gross total resection Follow-up at 6 months: pain resolved, constipation improving |
| 4 | M | 13 years | Yes, brother of Case 3 | Back pain, pelvic pain, and constipation (history of tethered cord and imperforate anus) | MR: partial agenesis of the sacrum below S2 and heterogeneous sacral mass and presacral mass (1.5 cm×2.0 cm×2.5 cm) | Combined anterior/posterior approach | Teratoma and ventral meningocele | Gross total resection Follow-up at 6 months: pain resolved, constipation improving |

Posterior approach

Midline dissection was carried down to the sacrum where multiple sacral laminectomies were performed. A midline durotomy was created inferior to the level of the most caudal exiting nerve root. Detethering was achieved by sectioning the filum terminale. Next, the thecal sac was circumferentially dissected, allowing for identification of the fistula, leading to the anterior sacral meningocele. The dissection was then extended below the coccyx into the presacral space, where it was ventrally mobilized from the posterior sacrospinous ligaments and ultimately fully resected.

DISCUSSION

Patients with CS most commonly present with severe constipation after birth.^[5,8] This is often due to an enlarging presacral mass. It most commonly includes an anterior sacral meningocele, enterogenous cyst, tethered cords, and/or teratomas (e.g. found approximately 50% of the time, with the potential for malignant transformation).^[1]

The female-to-male ratio of CS is 2:1 in children and 6:1 in adults. The increased female predominance in adults may be partially due to the frequency of CS diagnosed for patients presenting with dysmenorrhea (e.g., patient 2). About >50% of patients with CS have a mutation of HLXB9 located at 7q36; such mutations have been observed in up to 90% of familial-associated CS.^[4]

Utilizing a comprehensive, multidisciplinary team (oncology, pediatric surgery, and pediatric neurosurgery) and two-staged anterior/posterior surgical approaches, four patients with CS successfully underwent total resection of these lesions without any adverse events. We advocate early operative intervention for multiple reasons. First, symptoms can be improved with surgery. Second, early surgical intervention limits the risk of future, spontaneous rupture of the teratoma, resulting in meningitis, spinal abscesses, and/or malignant transformation. Third, surgical resection provides an accurate, confirmatory pathologic diagnosis.^[6]

CONCLUSION

CS is a rare condition associated with a triad of findings. Symptomatic sacral masses should be promptly treated with

anterior followed by posterior surgical resection. This provides an immediate pathologic diagnosis, while gross total resection can be safely accomplished without significant neurological injury. Alternatively, waiting and watching may result in not only a delay in diagnosis and the risk of malignant transformation but also increases the risk of permanent neurological injury.

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Conflicts of interest

There are no conflicts of interest.

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