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Case Report

Multidisciplinary surgical treatment of presacral meningocele and teratoma in an adult with Currarino triad

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Abstract

Background: Currarino syndrome (CS) is a rare genetic condition that presents with the defining triad of anorectal malformations, sacral bone deformations, and presacral masses, which may include teratoma. Neurosurgeons are involved in the surgical treatment of anterior meningoceles, which are often associated with this condition. The accepted surgical treatment is a staged anterior-posterior resection of the presacral mass and obliteration of the anterior meningocele.

Case Description: This case involved a 36-year-old female who presented with late onset of symptoms attributed to CS (e.g., presacral mass, anterior sacral meningocele, and sacral agenesis). She successfully underwent multidisciplinary single-stage approach for treatment of the anterior sacral meningocele and resection of the presacral mass. This required obliteration of the meningocele and closure of the dural defect. One year later, her meningocele had fully resolved.

Conclusion: While late presentations with CS are rare, early detection and multidisciplinary treatment including single-state anterior may be successful for managing these patients.

Key Words: Currarino syndrome, meningocele, sacral, teratoma

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INTRODUCTION

Currarino syndrome (CS) is an autosomal dominant syndrome. The classical presentation is characterized by sacral agenesis, anorectal malformations, and a presacral mass. CS is rare in adults, but should be recognized early to avoid life threatening complications, e.g. meningitis, rectal fistulas, and an approximately 1% risk for malignant transformation.

There are multiple surgical approaches utilized to treat anterior sacral meningoceles (ASM) and presacral masses. The most common is a two-staged anterior-posterior approach that carries an increased risk of anorectal perforation leading to infection along with greater morbidity attributed to increases blood loss, operating time, and length of hospital stay. Here, we offer a single-staged approach wherein the anterior sacral meningocele and presacral mass were both treated in one sitting.[1,3]

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CASE REPORT

A 36-year-old female presented with intermittent constipation and bilateral lower extremity radiculopathy in the L4 distribution on the right and L5 distribution on the left. Family history was significant for the diagnosis of CS in both her children. The magnetic resonance imaging (MRI) and computed tomography (CT) scans revealed an incidental anterior sacral meningocele, a perirectal mass, with sacral agenesis, as well as right hemisacral hypoplasia, and an 8.0 cm × 8.0 cm × 6.0 cm meningocele emerging from a ventral sacral defect [Figures 1-3]. A 7.5 cm × 5.0 cm cystic heterogeneously enhancing mass was also noted immediately inferior to the meningocele, adjacent to the sigmoid colon. The remainder of the neuraxis appeared normal.

Surgery

Surgical treatment of the meningocele required a complete S3-4 laminectomy with transdural closure and obliteration of the meningocele pedicle. The laminectomy, along with an extended incision to the anal sphincter, provided excellent exposure of the presacral mass. With the assistance of general surgery, the presacral mass was grossly excised without complications. Pathologically, this proved to be a mature teratoma. One year later, the myelomeningocele had fully resolved [Figure 4].

DISCUSSION

CS is a caudal regression syndrome typically associated with the classic triad of sacral malformations (sickle shaped sacrum, sacral agenesis below S2 [Figure 5], and rarely complex malformations), anorectal malformations (anal atresia, low and high anal imperforation/rectal fistulae), Hirschprungs disease, and presacral masses (anterior myelomeningocele, malignant or benign teratoma, dermoid cyst, epidermoid cyst, leomyomasarcoma, lipoma, or a combination thereof).[2,5,7]

Genetic discussion

This phenotype has been linked to loss of function mutations that disrupt the HLXB9 homeobox gene

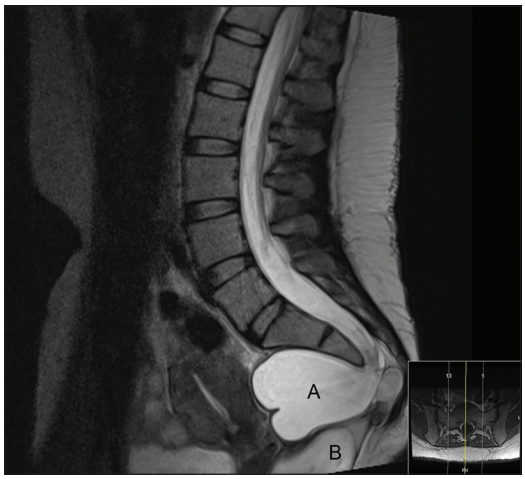

Figure 1: Sagittal MRI, preoperative. (A) Anterior sacral meningocele; (B) teratoma

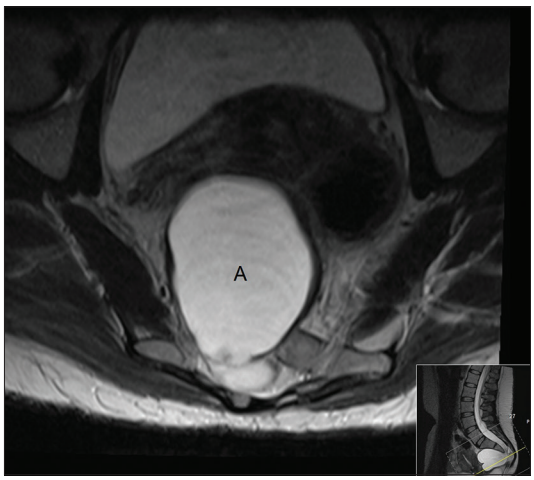

Figure 2: Axial MRI, preoperative. (A) Anterior sacral meningocele

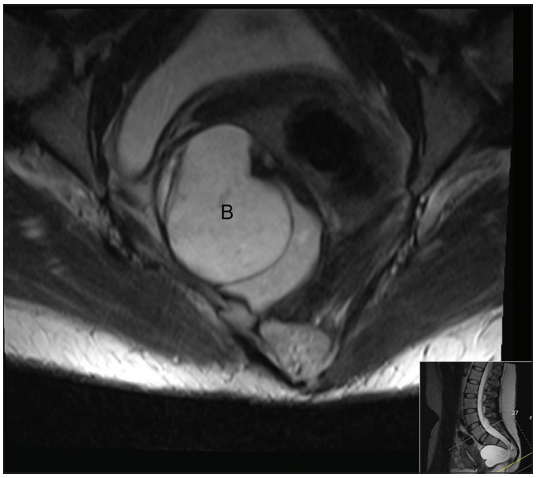

Figure 3: Axial MRI, preoperative. (B) Teratoma

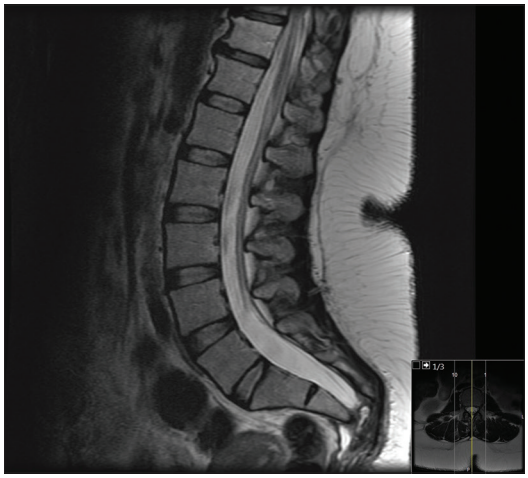

Figure 4: Sagittal MRI, postoperative

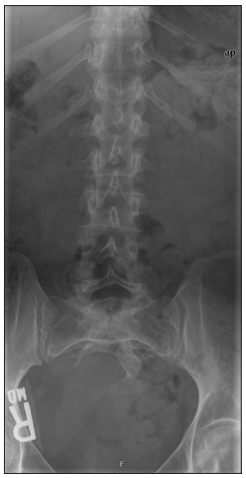

Figure 5: Coronal XR, preoperative, showing sacral agenesis

located on chromosome 7q36, and which encodes the HB9 nuclear protein. Although it is most commonly associated with nonsense mutations, a variety of mutations have been described including missense, splice site, and frameshift.[4,5] The disease process itself follows an autosomal dominant inheritance pattern with variable penetrance and weak phenotype-genotype correlation.[4,8]

Clinical diagnoses

More than 80% of the patients with the classic triad are diagnosed within the first decade of life.[2] This patient presented at the age of 36 with radiculopathy, and was older than those typically diagnosed with CS. A review of 205 patients by Lynch *et al*. showed that renal/urinary symptoms were among the three common defining features of CS.[8] Furthermore, one-third of patients were asymptomatic at the time of diagnosis (incidental finding). A very common presenting symptom was chronic constipation in childhood attributed to the ventral meningocele and/or pre sacral masses, dysganglionosis due to malformations of the enteric nervous system (i.e., Hirschprung's), tethered cords, or errors in migration of the pluripotent cells of the caudal eminence after primary neurulation.[2,6,8]

Surgical approaches

Classic CS is routinely treated in a two-staged approach with the anorectal malformation and presacral mass dealt separately from the obliteration of the anterior meningocele.[2] Here, we opted for a single-stage operation completing simultaneous excision of the mass and ligation of the ASM, thereby avoiding infection and a need for a second operation. In addition, this single staged posterior approach reduced the need for a colostomy, which further reduced the risk of infection.

CONCLUSION

While late presentation of CS is rare, early detection and multidisciplinary treatment including surgical resection is critical for the successful management of these patients because these lesions may undergo malignant transformation. Depending on the lesions present, a single-stage approach minimizes surgery and reduces its inherent risks including those of postoperative infection.

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Conflicts of interest

There are no conflicts of interest.

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