Posterior Sagittal Approach for Currarino Syndrome With Anterior Sacral Meningocele: A Case Report

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The authors report a case of Currarino syndrome with anterior sacral meningocele, tethered cord, and anorectal stenosis that was treated by posterior sagittal approach. Initially, a diverting colostomy was performed. Two months later, excision of the meningocele, untethering of spinal cord, and anorectoplasty were performed simultaneously without complication. Posterior sagittal approach seems to be very

CURRARINO SYNDROME is a rare condition characterized by the triad of anorectal malformation, sacral bony defect, and presacral mass. It was first described by Currarino et al¹ in 1981 to be a result of abnormal separation of the neuroectoderm from the endoderm. In this syndrome, anterior sacral meningocele is one of the most common presacral masses to be reported.² Cases involving spinal connection require management to avoid infectious complications during surgical treatment. We describe a case of Currarino syndrome with anterior sacral meningocele that was treated by posterior sagittal approach (PSA) after a preliminary colostomy.

CASE REPORT

A 9-month-old girl was referred to our clinic with a complaint of severe constipation since 5 months of age. Her anus was so deep and narrow that even the small finger of the examiner could not be passed on digital examination. A plain pelvic radiograph found sacral scimitar bony defect, and severe anorectal stenosis was determined by subsequent barium enema study. Anorectal manometric study showed high anal pressure but normal anorectal reflex. After the diagnosis of congenital anorectal stenosis, she underwent sigmoid colostomy. Thereafter, computed tomography (CT; Fig 1) and a magnetic resonance imaging (MRI; Fig 2) study showed a presacral mass, which was presumed to be an anterior sacral meningocele with tethered spinal cord. She was diagnosed as having a variant of Currarino syndrome. At age of 11 months, she underwent simultaneous operation by posterior sagittal approach (PSA) and laminotomy. By PSA, the anorectal tube was freed from the meningocele, which encircled about three fourths of

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Copyright © 2000 by W.B. Saunders Company 0022-3468/00/3507-0021\$03.00/0 doi:10.1053/js.2000.7838 beneficial for excision of the presacral mass and reconstruction of the anus.

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the rectum. The dissection continued to the anterior surface of the presacral mass. The neurosurgeon performed laminotomies of S2 and S3 vertebrae and freed the filum terminale aided by microscopic visualization. The defect of the dura mater was repaired with Gore-Tex Tissue Patch (W.L. Gore and Associates, Flagstaff, AZ). Total excision of the presacral mass and anorectoplasty were performed after resection of 3 cm of fibrous stenotic anorectum (Fig 3). Pathological finding of the resected tumor showed a lipomeningocele and a dermoid cyst.

Postoperative course was excellent without any complications such as meningitis. The colostomy was closed after 3 months. Two years





Fig 1. CT showed a presacral mass that was presumed to be an anterior sacral meningocele. The rectal wall was thickened and displaced ventrally by presacral mass. A silicon catheter was inserted into the rectum.

postoperatively, she is doing well without any recurrence of presacral mass or retethering of spinal cord. She is continent and shows no signs of constipation.

DISCUSSION

The most frequent symptom of Currarino syndrome is constipation as seen in this case. Urinary tract problems, meningitis, and perianal infections also are reported.¹⁻⁸ Our patient had a pelvic radiograph that showed scimitar sacral bony defect. The pelvic MRI was especially useful in differentiating the presacral mass and detecting tethered spinal cord.3-5,9 In most cases of Currarino syndrome, excision of presacral mass and anorectoplasty are required. In cases of tethered spinal cord, release of spinal cord also is required. In surgical treatment of cases with anterior meningocele, some investigators recommended a staged operation including initial excision of meningocele and latter anorectoplasty to avoid infectious complications such as meningitis.^{3,6} But in this case, we performed these procedures simultaneously without any infectious complications. Diverting colostomy and strictly planned preparations were essential to avoid infection. In staged operation, anorectoplasty may be more difficult because of postoperative adhesions and scarring after



Fig 2. Sagittal section of MRI showed an anterior sacral meningocele and tethered spinal cord. (A) T1-weighted image. (B) T2weighted image.



Fig 3. The anterior sacral meningocele was resected, and the rectum was freed completely from presacral mass. Dura mater was repaired with Gore-Tex Tissue Patch (W.L. Gore and Associates) after untethering of spinal cord.

initial operation for meningocele and tethered spinal cord. The rectum can, on occasion, be encircled by presacral mass in addition to causing extrinsic compression.^{1,3,7,8} Simultaneous operation by PSA and spinal surgery is beneficial in achieving complete excision of the presacral mass, untethering of the spinal cord, and easier reconstruction of the anorectum.

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