

The Shrinking of an Anterior Sacral Meningocele in Time Following Transdural Ligation of its Neck in a Case of the Currarino Triad

Currarino Triadı Olgusunda Anterior Meningosel Kesesinin Boynunun Bağlanmasıyla Kesenin Zamanla Küçülmesi

ABSTRACT

In this paper we present an adult patient with the Currarino triad, associated with six habitual abortuses and chronic constipation. Our aim is to report the effectiveness of a simple surgical technique in the treatment of anterior sacral meningocele in the Currarino Syndrome. The presentation of the Currarino Syndrome in adulthood is extremely uncommon. We performed a posterior procedure via lumbar and sacral partial laminectomy and transdural ligation of the neck of the meningocele mass. There were no additional neurological deficits and no recurrence of the presacral mass over the 2-year follow up period. When an adult female with habitual abortus and chronic constipation is seen, the possible presence of the Currarino Triad should be investigated and treated. The anterior sacral meningocele in a case of the Currarino Triad regresses over time following transdural ligation of its neck.

KEY WORDS: Anorectal stenosis, Anterior sacral meningocele, Currarino triad, Currarino syndrome, Habitual abortus

ÖZ

Bu makalede 6 habitüel abortus ve konstipasyon öyküsü olan Currarino Triadlı bir erişkin hastayı sunuyoruz. Amacımız Currarino sendromunda anterior sakral meningoselin tedavisinde basit bir cerrahi tekniğin etkili olabildiğini göstermektir. Currarino sendromunun erişkinde ortaya çıkması son derece nadirdir. Posterior bir girişimle lomber ve sakral kısmi laminektomi yaparak meningosel kesesinin boynunu transdural bağladık. Postop ek nörolojik defisit gelişmedi ve iki yıllık takip süresi boyunca presakral kitle nüksetmedi. Habitüel abortus ve kronik konstipasyon öyküsü olan bir hastada Currarino sendromu olasılığı araştırılmalı ve tedavi edilmelidir. Currarino triadında anterior sakral meningosel, boynunun transdural bağlanmasıyla zamanla geriler.

ANAHTAR SÖZCÜKLER: Anorektal stenoz, Anterior sakral meningosel, Currarino triadı, Currarino sendromu, Habitüel abortus

Abbreviations used in this article: AM=anorectal malformation; ASM=anterior sacral meningocele; CT=computed tomography; MRI=magnetic resonance imaging

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INTRODUCTION

The Currarino triad, also known as the “Currarino Syndrome”, is a rare complex of congenital caudal anomalies including three main features; a sacral bony deformity, anorectal malformations (ARM), and a presacral mass (2, 5). Enteric cysts, dermoid cysts, lipomas, leiomyosarcomas, yolk sac tumors, pelvic hamartomas and carcinoid tumors may all be described as pre-sacral masses. However, the most frequent pathologies described as such are anterior sacral meningoceles (ASMs) and teratomas (5).

Although Currarino described the triad as a syndrome complex in 1981, it was first reported by Kennedy in 1926 as an association of sacral bony defect, anterior sacral meningocele (ASM), and rectal polyp (5, 14). The Currarino triad is likely to represent only about 5% of symptomatic patients with anorectal malformations. The triad may be complete if all three anomalies are present, or incomplete if the triad is associated with only 1 or 2 of them (8, 29, 32). Approximately 250 cases have been reported to date in the medical literature (5, 7, 8, 10-12, 15-17, 20, 26, 28, 29, 32). In most cases, the syndrome is diagnosed in infancy or childhood. The presentation of the Currarino Syndrome in adulthood is extremely uncommon. Here we present an adult patient with the Currarino triad, associated with habitual abortus and chronic constipation.

CASE

A 30-year-old obese woman was admitted to the gynecology department in our hospital with a history of six habitual abortuses. She complained of low back pain for 6 years and chronic constipation since birth. An anal dilatation had been performed when she was 6 months old to treat anal stenosis. A gynecological work-up revealed uterus bicornis. A plain radiograph and pelvic computed tomography (CT) showed a scimitar-shaped sacrum (Figure 1). Pelvic magnetic resonance imaging (MRI) scan showed an extremely large ASM, a low-lying conus medullaris and a thickened filum associated with a lipoma (Figure 2). When she was referred to our department of neurosurgery, her neurological examination was normal.

An anterior laparoscopic procedure was performed first but we were prevented from reaching the pre-sacral region by an excess of fatty tissue. In the second operation, we performed a posterior approach via a lumbosacral partial



Figure 1: Plain radiograph shows a scimitar-shaped sacrum.



Figure 2: Pelvic MRI shows a very large ASM, a thickened filum and a low-lying conus medullaris associated with a lipoma.

laminectomy. The anterior sacral meningocele was exposed intradurally and extradurally. The tight filum terminale was severed in the same operation. Cerebrospinal fluid was aspirated from the cyst and the meningocele was ligated and cut at the neck. Post-operative MR scans obtained early after surgery and six months later showed the regression of the ASM (Figure 3 A and B). At two-years follow-up, neurological examination was normal. The patient was not complaining of constipation anymore.

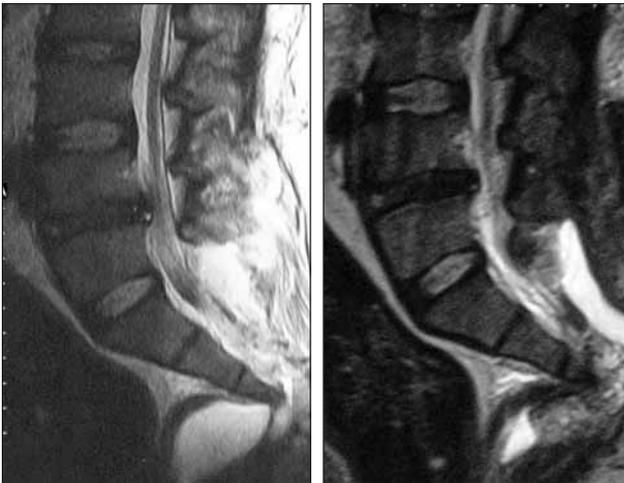


Figure 3: (A-B): The MR scans, obtained during the early post-operative period (A) and six months after surgery (B) show the regression of the ASM.

DISCUSSION

There is an increasing number of published reports about mutation analysis and the genetic factor in the Currarino Triad (9,11,16,20). It is an autosomal dominant hereditary syndrome and a responsible gene has been mapped at chromosome 7 (7q36) (6,7,16). This linkage has recently been refined to a homeobox gene, HLXB9, as the major gene for inherited sacral agenesis (9,20). Unfortunately, we were unable to obtain any genetic information about our patient.

The most common and the cardinal symptom of the Currarino triad is chronic constipation, as in this case. However, other presentations such as abdominal distention, bowel obstruction, recurrent meningitis, low back pain, headache and/or urinary tract infections may occur (8, 10, 28). Besides these symptoms; dysmenorrhea, obstructed labor, perianal sepsis may also constitute the initial complaints. The cause of constipation in the Currarino syndrome remains unknown (7). The resection of a presacral mass may not relieve patients from constipation (7). However, our patient was no longer complaining of constipation two years after the operation, hence the anterior sacral meningocele might have caused this symptom.

Additionally, our patient presented with habitual abortus. To our knowledge, there is no published reference to this kind of presentation of an adult case in the literature. Emans et al drew attention to the theoretical possibility of abortus, even though one of

their patients with a large anterior meningocele had an uneventful pregnancy and vaginal delivery (7). There may be other reasons for abortus in our patient. Pregnant women are prone to becoming folate deficient as the folate intake of pregnant women is often insufficient. A low maternal folate status may result in abortus (23). The role of folic acid in the etiology of neural tube defects is well known. Because the patient had an anterior sacral meningocele and experienced multiple abortus, it was possible that we missed a familial metabolic disorder to diagnose. However, the patient stated that she had vitamin supplements during the pregnancies. Complex congenital anomalies such as uterus bicornis, unicornuate uterus and uterus didelphys may negatively affect fertility (1, 24, 30). Anatomic uterine defects appear to predispose women to reproductive difficulties, including first- and second-trimester pregnancy losses, higher rates of preterm labor and birth, and abnormal fetal presentation (24). Therefore, the most probable reason of habitual abortus seen in our patient was uterus bicornis, not the anterior sacral meningocele.

The female-to-male ratio has been reported to be 3:1 in nonfamilial cases and 1:1 in familial cases (8). In more than three quarters of the reported patients, some bony abnormality of the anterior sacral elements was present, usually a "scimitar-shaped sacrum" (absence of part of the sacrum with preservation of the first sacral vertebra and a remnant of S2–S5), as seen in this case (3, 22, 27). This sacral defect is pathognomonic for this triad. All first-degree relatives of the patient should be offered a pelvic x-ray examination (11,16). We found no similar case in the family pedigree of our patient. Anorectal malformations (ARMs) appear most frequently as anorectal stenosis, as seen in our patient. Rectal stenosis, anal ectopia and atresia are the other ARM pathologies (8,16,20). The incidence of the Currarino Triad is high in patients with anal stenosis and it should be suspected in such cases. Pelvic ultrasonography, CT scanning and MRI are all useful diagnostic tools (3,12,26,27). However, MRI allows the early diagnosis and precise anatomical definition of the pre-sacral mass (3,22). In our patient, MRI scans showed the intensity characteristic of cerebrospinal fluid in the cystic pre-sacral mass. MRI can also determine further associated malformations such as tethered cord, holocord syringomyelia, intraspinal lipomas,

rectovaginal fistulas and partial duplication of the vagina, uterus or urinary tract (3,22,31).

Surgical techniques in the treatment of the Currarino Triad

Early diagnosis and treatment are essential to avoid morbidity and mortality (18). In routine practice, the anorectal malformation should be treated first. If this is not severe, conservative management can be performed (12,19,25). Radical surgery should be undertaken as soon as an anterior sacral meningocele is found and the anorectal malformation treated later as rupture of a meningocele can occur spontaneously, and may result in meningitis, (12,17,19,28). The goal of ASM treatment is to eliminate contact between fluids in the spinal subarachnoid space and the meningocele (13,19).

One surgical technique for treating ASM is the posterior sagittal approach, which involves complete exposure of the perineal region by means of a median sagittal incision running from the sacrum to the anal region (19,21). Another surgical approach is via a posterior lumbar and/or sacral laminectomy and transdural ligation of the neck of the meningocele. This procedure is ideal when the stalk is narrow (12,15,19). We performed a posterior approach via a laminectomy-laminoplasty procedure on our patient. This approach provides better exposure of the pedicle, better protection of the sacral nerve roots and watertight closure of the meningocele pedicle. The latter is essential to prevent recurrence. Because of the usual close adherence of the sac to the posterior wall of the rectum, an extensive resection of the tumor capsule or meningocele membrane at surgery should be avoided and a limited sac removal is recommended (13,19). Resection of a presacral meningocele is unnecessary after closure of the connection with the thecal sac. The residual meningocele sac will be absorbed gradually over time.

Endoscopic treatment for ASM (the posterior or anterior endoscopic approach) is another reported alternative method (4,13,25). There are two successful posterior and one successful anterior endoscopic treatments of ASM as the initial treatment method in the literature (4,13,25). We also tried an anterior endoscopic method in our patient, but we were unable to reach the meningocele pedicle because of fatty tissues. This method can be an alternative surgical therapy in patients who are not obese.

The incidence of the association between the Currarino triad and tethered cord syndrome is high and spinal cord untethering is also indicated (32). Sometimes a few sacral roots protrude into the sac and may be adherent to the meningocele wall. It is necessary for neural structures to be carefully dissected free and retrieved back into the normal thecal space before ligation of the meningocele neck (17,32). Some authors advocate that there is no indication for routine tethered cord release in the Currarino triad (7). However, advising against surgery when tethering is present is not the majority view.

In conclusion, although rare, the possibility of the Currarino Triad should be investigated and treated when an adult female with habitual abortus and chronic constipation is seen. The anterior sacral meningocele in the Currarino Triad regresses over time following transdural ligation of its neck.

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