

Currarino Triad

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The authors report a case of Currarino triad which had a congenital anorectal stenosis associated with a sacral defect and a presacral mass. A 1-year-old female presented with constipation since birth. Neurological deficits were not found on admission. She had had a diverting colostomy due to anorectal stenosis at another hospital before admission. Lumbar X-ray films showed bony defect caudal to the third sacral vertebra. Magnetic resonance image demonstrated a round cystic pelvic mass which was connected with a dural sac via anterior sacral defect. Posterior approach with Intradural removal of the presacral cystic mass was performed and followed by anoplasty by a pediatric surgeon. The cystic mass was verified histologically as mature teratoma with cystic change. Postoperatively, the urinary function and bowel movement remained intact. Currarino triad should be suspected and evaluated physically and radiographically in a case of congenital anorectal stenosis. Prompt recognition and close cooperation between pediatric surgeons and neurosurgeons is advisable to ensure adequate surgical treatment.

KEY WORDS : Currarino triad · Presacral mass · Mature teratoma.

Introduction

The Currarino triad^{5,6)} was first described by Currarino et al in 1981. This triad consists of an anorectal malformation, sacral bony defect, and presacral mass^{5,6)}. The congenital malformation is known to be presumably caused by abnormal separation of the neuroectoderm from the endoderm⁵⁾. The problems with this triad are difficulties in early diagnosis and surgical management. Therefore, suspicion of this anomaly in congenital anorectal stenosis is crucial for proper diagnosis and treatment¹⁰⁾. We experienced a rare case of Currarino triad and report it.

Case

History

A 11-month-old female with presacral cystic mass was admitted at another institution because of constipation and progressive abdominal distension since birth. She had colostomy which was performed at age 30 days in another institution because of an anorectal stenosis. Incidental presacral mass was found on lumbar computed tomography scan. She was trans-

ferred to our institution for the further evaluation. She had no familial history.

Physical examination showed colostomy on the left lower abdominal area. Index finger was not introduced on rectal examination. Motor and sensory system was intact. Peripheral blood examination, chemical analysis, and routine urinalysis were normal.

Pelvic films showed a defect of the lower part of the sacrum (Fig. 1A). Barium enema examination confirmed a severe anorectal stenosis and anterior displacement of the rectum (Fig. 1B). Lumbar magnetic resonance imaging showed a 3 × 3cm presacral cystic mass before the 3rd sacrum (Fig. 2A). Repeated enema was initiated for preop rectal emptying. We consulted to pediatric surgeon for co-operation.

Operative finding

The midline skin incision, subperiosteal dissection, and the laminectomy was made from the 2nd to 4th distal sacrum. The dura and arachnoid were opened in the midline. At that time, the opening, through which the cystic mass was communicated with the dural sac, was identified 0.5cm superior to the end of the filum terminale. The cystic mass contained clear cerebrospinal fluid within a thin wall. The fluid was aspirated through the opening. The cystic mass was excised completely through it. Nerve roots or other abnormal tissue were not present in it. The opening was intradurally closed with 5-0 black silk. Dura mater, subcutaneous tissue, and skin was approximated. After the previous skin incision was covered with aseptic plastic

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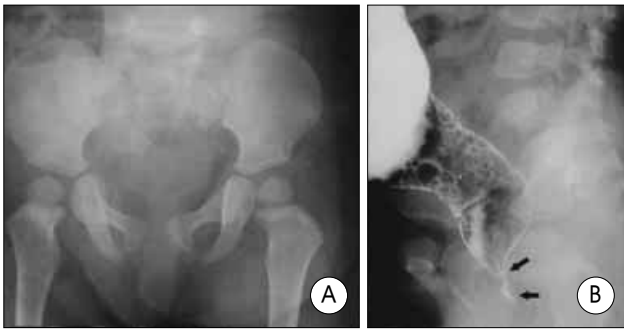


Fig. 1. A : Plain radiograph of the bony pelvis shows a small defect at the lower sacrum. B : Barium enema examination of patient reveals anorectal stenosis and an anterior displacement (arrows) of the rectum.



Fig. 2. Sagittal T2-weighted magnetic resonance image showing a round cystic presacral mass (A) with homogeneous content of high signal intensity (arrow) which is removed postoperatively (B).

coverage, the new incision was extended to the posterior portion of the anus and anoplasty was made by a pediatric surgeon.

Pathologic finding

Grossly, an irregular shaped grayish white solid mass measured 1.5 × 1.2 × 1.0cm. Histologically, it was composed of stratified squamous epithelium, fibrous connective tissue, fat tissue, and ependymal cells. Also, it was containing an admixture of glial tissues (Fig. 3). Its diagnosis was mature teratoma with cystic change.

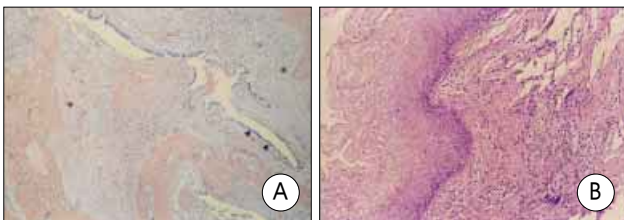


Fig. 3. Microscopic finding. A : Large amount of glial tissue (asterisks) and space lined by ependymal cells (arrowheads) surrounding the collagenous tissue (H & E X 40). B : Stratified squamous epithelium and upper dermis (H & E X 400).

Postoperative course

The urinary function and bowel movement were intact. Colostomy was closed 1 month later. MRI showed removed status of the cystic mass at 1 month postoperatively. She had no evidence of recurrence at 2 year follow-up period.

Discussion

The association of rectal stenosis, presacral mass and sacral deformity was described by Currarino et al. in 1981^{6,10,12,15}. Currarino reported three infants in whom a congenital anorectal stenosis was associated with a sacral defect and a presacral mass was treated⁵. The three cases suggest a specific malformation complex characterized by three main features : (1) a congenital anorectal stenosis or another type of low anorectal malformation; (2) an anterior sacral defect; and (3) a presacral mass which may be a meningocele, a teratoma, occasionally an enteric cyst, or a combination of these. Presacral tumors are usually located in the potential space between the perirectal fascia and the fibrous coverings of the anterior sacrum⁶. Our case had all three features; presacral mature teratoma, sacral defect, and anorectal stenosis.

The embryogenesis of the malformation complex is not certain. Abnormal endoectodermal adhesions and notochordal defects in very early fetal life have been postulated as the underlying embryologic cause for these lesions by Currarino et al^{5,6}. Before the 20th day a close relation exists between endoderm(future intestinal tract) and ectoderm(future spinal column). Mesodermal tissue then forms laterally and fuses in the midline around the notochord to form the vertebral column, thus separating endoderm and ectoderm. Failure of this mesodermal fusion, either due to adhesions or to anomalies of the notochord, results in persisting connections between intestinal tract and spinal column in the form of a fistula, meningocele, enteric cyst or teratoma⁶. The anorectal anomaly is a consequence of these early abnormal adhesions between the hind gut and the neural tube. The presacral teratoma is formed by local enteric and neuroectodermal elements combined with mesodermal elements brought into the space by the developing somites and other local mesodermal structures. This is called "the split notochord syndrome"⁶.

In Currarino's triad, most of the symptoms resulted from compression or obstruction of adjacent organs or from pressure on pelvic nerves or bone. Among these patients, constipation is the most common presenting symptom³. Especially, when severe constipation since birth was presented, these associative anomaly should be evaluated. Symptoms are characteristically

present for many months or years, and tumors may obtain considerable size, although they are palpable by digital examination of the rectum⁹. The age at diagnosis were mostly less than 12 years of age¹⁵. Females were more commonly affected (female : male=30 : 21). Our case had constipation at birth. Many report had familial tendency^{12,15,17}, but our case had not. Lee et al¹⁰ in Seoul university reported 11 cases of Currarino triad at 1997 and presacral masses included 7 teratomas, 2 meningoceles, 1 dermoid cyst, and 1 enteric cyst with dermoid cyst. Heji reported of 4 cases of Currarino triad that included 2 teratomas, 1 hamartoma, and 1 not confirmed. Park et al¹² reported 4 cases : anterior meningocele and lipoma, dermoid cyst : anterior meningocele : enterocele, dermoid : dermoid cyst.

The early recognition and treatment of correctable lesions of the terminal spinal cord in patients with anorectal malformations may preserve important neurologic function^{1,7}. When congenital rectal stenosis is detected in the newborn during the initial physical examination, the presence of congenital rectal stenosis in infancy should raise the examiner's index of suspicion to evaluate the patient for possible presacral abnormalities. Because presacral teratoma, anterior sacral meningocele, or bony anomalies may be the underlying extrinsic causes of congenital rectal stenosis¹¹. Therefore, rectal examination of all patients with severe constipation is mandatory to detect rectal stenosis, which is always located distally in this syndrome.

Radiographic examination of the bony pelvis in these patients may reveal sacral defects. Barium enema is likely to demonstrate anterior displacement and poor emptying of the rectum. Barium enema should be the initial step in identifying the mass and the extent of the anorectal stenosis. In our case, rectum was displaced anteriorly and narrowed, through which dye had not passed. Myelography helps to find the evidence of spinal-abdominal communication^{2,13}. Appropriate examination of the retrorectal space and the lumbosacral spine for potential anomalies is strongly indicated. Pelvic ultrasonography, computed tomography scan or magnetic resonance imaging can be performed to detect, differentiate, and identify the extent of the presacral mass. Computed tomography scan has proved valuable in determining the extent and degree of tumor invasion. Magnetic resonance imaging is the diagnostic procedure of choice because its multiplanar imaging capability is quite useful in determining the connection of the presacral mass to the thecal sac². In our case, MRI showed that a large round mass with homogenous content of high signal intensity at T2-weighted sagittal image, which was connected with the thecal sac.

Proper management in each case should be based on the

individual variations of the triad. Most cases were treated successfully via a sacral approach (posterior sagittal approach). Important factors in the selection of operation include size and location of the pelvic mass and myelographic data such as the size and level of the communicating neck and any deformity or malposition of the spinal cord or cauda equina. The choice will also vary with the surgeon's experience². Stewart, et al described that for lesions <10cm in size a posterior parasacral approach is often feasible but for larger lesions an abdominal approach or combined approach should be considered¹⁴. Anterior approach is preferred in case of a large presacral mass. Some cases is required both an abdominal and posterior approach to remove the tumor^{3,10}.

The management of these patients begins with establishing the true nature of the presacral mass. Complete removal of the mass is the object of treatment. The need to excise a presacral teratoma is based on two considerations : first, that malignant presacral teratomas have been reported. Second, the presence of teratoma inside the rectal wall in a number patients. The single most reliable indicator of malignancy is the presence of pain or destruction of the sacrum; however, large benign tumors can present a similar pattern⁹. But, removal of presacral mass is difficult because the mass is firmly attached to the posterior wall of the rectum and dura¹⁵. The surgeon should bear in mind that the mass is firmly attached to the posterior wall of the rectum and/or dura to avoid rectal perforation during the dissection. In our case, when the content of cystic mass was aspirated through the opening, fortunately the cystic wall was easily dissected from the posterior wall of the rectum and could be removed completely. Confirmatory pathology was mature teratoma with cystic change, without any nervous tissues in it's contents.

In most severe rectal stenosis diverting colostomy and bowel preparation will be indicated because of massive fecal stagnation^{6,8}. When the presacral mass is removed, repair of this lesion is performed initially and anorectal malformation should be treated later by staged operation because there is a significant risk of meningitis with a simultaneous operation¹⁰. Neurosurgical exploration should precede intestinal surgery⁶. Associated anorectal fistula should be evaluated preoperatively to decrease risk of infection. Postoperative abscesses or meningitis are not uncommon and these infections can occur preoperatively due to preexisting unrecognized rectal fistula, or to infection of enteric cysts^{5,8}. In our case, preoperative diverting colostomy and barium enema was helpful to evaluate the presence of the rectal fistula.

If a sacral bony defect is present, computerized tomography scan is suggested. This sacral defect gives occasionally the

routes for approach to the presacral mass. Sacral defect is not recommended for correction.

Reported complications include meningitis, ruptured meningoceles, retrorectal abscesses, and a low incidence of malignancy of the presacral teratoma¹⁷⁾. Also, other complications reported included obstruction of labor with fetal or maternal death and several instances of lethal meningitis after operation^{2,17)}. Because of the potential for infection, needle aspiration is not recommended^{4,9)}. Any surgical procedure involving the presacral area has a chance of injury to the pudendal nerve and the upper three sacral nerves to cause neurogenic bladder.

Conclusion

We experience a rare case of Currarino triad. Prompt recognition and appropriate operative management of the presacral lesion with Currarino triad can relieve obstructive symptoms and minimize morbidity. Close cooperation between pediatric surgeons and neurosurgeons is advisable to ensure adequate surgical treatment, considering the risk of rectal perforation and intraoperative nerve damage.

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