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Samer S. Hoz

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Incomplete Currarino triad in an adult woman. A case report

Rasha A. Al-Youzbaki[1], Emad Hazim Mahmoud[1], Zahraa A. Alsubaihawi[2], Mohammed A. Almeran[1], Saja A. Albanaa[2], Samer S. Hoz[3]

[1] Department of Neurosurgery, Ibn Sina Teaching Hospital, Mosul, IRAQ
[2] College of Medicine, Baghdad University, IRAQ
[3] Department of Neurosurgery, University of Cincinnati, Ohio, USA

ABSTRACT

Introduction: Currarino Syndrome (CS) is a rare entity characterized by a triad of sacral agenesis, anorectal malformations, and pre-sacral masses. CS is typically diagnosed during the first decade of life.

Case Description: We present a rare case of incomplete Currarino syndrome manifesting in a 36-year-old lady who presented with back pain, urinary retention, anal paresthesia, and lower limb weakness. The patient underwent multiple laminectomies and partial resection of an epidermoid cyst and regained function.

Conclusion: Although rare, the possibility of Currarino syndrome should be entertained in adult patients with lower lumbosacral symptoms and a pre-sacral mass. A thorough physical examination and strategic pre-operative planning are mandatory to maximize patient outcomes.

Keywords
Currarino syndrome, sacral agenesis, epidermoid cyst

Corresponding author:
Saja Albanaa

University of Baghdad, College of Medicine, Iraq

sajaalbanaa@gmail.com

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INTRODUCTION

Currarino syndrome is a rare entity with a set of complex congenital anomalies. It manifests clinically as a triad of sacral bony defects, anorectal malformations, and pre-sacral masses, such as anterior meningocele (1,2). This condition is usually diagnosed during the first decade of life and diagnosis in adults is rare (3,4). Both sporadic and inherited cases have been reported, with an estimated incidence of 1 in 100,000 of the population (5,6). Here, we report a rare case of an incomplete Currarino syndrome (sacral agenesis, epidermoid cyst with no anorectal malformations) manifesting in adulthood.

CASE DESCRIPTION

A 36-year-old female presented with a two-week history of lower back pain, urinary retention, and anal numbness. She had no history of trauma. Her examination was significant for sacral anesthesia and weakness in ankle plantar and dorsiflexion -Medical Research Council (MRC) scale 4/5 -. Magnetic resonance imaging (MRI) of the lumbosacral

spine and pelvis revealed sacral agenesis and an intradural cystic lesion extending from the level of the second lumbar vertebrae (L2) to the sacrum with an anterior meningocele (Figure 1).

Multiple laminectomies (from L2 to sacrum) were performed through the posterior approach. . The cystic lesion was identified and durotomy revealed a soft white pearly lesion suggestive of epidermoid tumor which was partially resected as it wrapped most nerve roots with an adherent surrounding capsule. The histopathological analysis revealed flakes of keratinous materials with strips of unremarkable squamous epithelium consistent with epidermoid cyst (Figure 2).

The patient had an uneventful recovery and was discharged on a course of steroids and referred to a bladder training program. On her one-month follow-up, her bladder function was back to normal as were her lower limb power and peri-anal sensation. The MRI showed a decrease in the size of the epidermoid cysts and anterior meningocele (Figure 3).

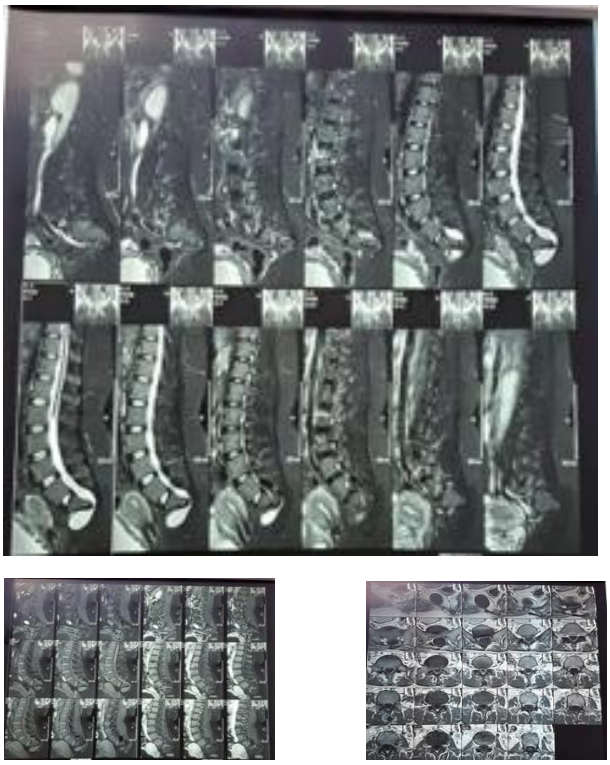

Figure 1. A, B MRI sagittal view of the lumbosacral spine T1 and T2 showing an intradural mass hypointense in T1 and hyperintense in T2 extending from L2 to the sacrum and turning anteriorly to the. Partial agenesis of the sacrum is also evident. C: MRI axial view showing cystic cavity in the sacral spine extending anteriorly in the pelvis through a widened sacral foramen.

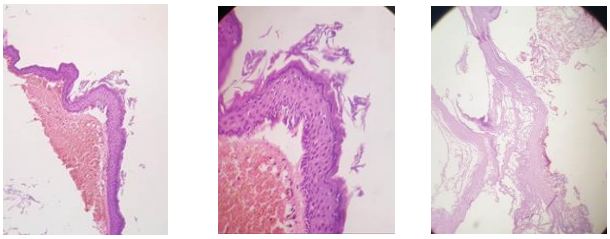

Figure 2. Pictures of histopathology showing flakes of keratinous materials with strips of unremarkable squamous epithelium consistent with epidermoid cyst.

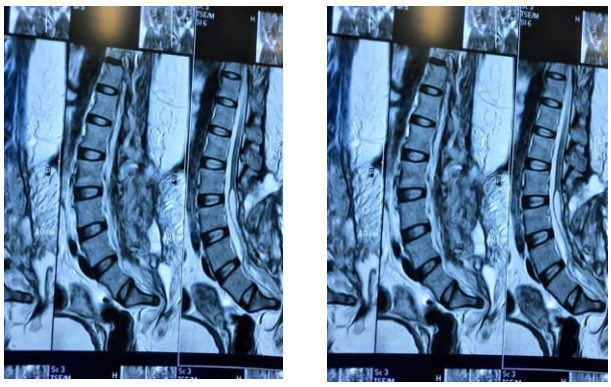

Figure 3. Post-operative MRI T1 and T2 sagittal view showing operative site from L2 to the sacrum and decrease in the size of the intradural lesion and the anterior meningocele.

Discussion

Sacral agenesis is a congenital deformity that ranges from partial agenesis of the unilateral sacrum to complete bilateral sacral agenesis (7-9). In a normal pregnancy, the incidence of sacral agenesis is between 0.005 and 0.1%. However, 16-20% of sacral agenesis patients have diabetic mothers. Sacral agenesis is associated with an increased incidence of spinal dysraphism, and currarino triad. (10)

Spinal meningocele is a posterior protrusion of meningeal elements out of the spinal canal, usually seen in the lumbosacral region. An anterior sacral meningocele is a meningeal protrusion into the presacral, retroperitoneal, and intraperitoneal space as a consequence of partial anterior sacral agenesis (11). Anterior sacral meningoceles may occur anteriorly through a defect in the body of sacrum or anterolaterally through an enlarged intervertebral foramen or coalesced foramina as in our case (12) Such improper development can result from other coexisting abnormalities in the skin, subcutaneous tissues, spine, and internal organs. Anterior sacral meningocele occurs sporadically, but familial cases have been reported in literature as part of the Currarino triad. (13)

The most common presacral masses are teratomas and anterior sacral meningocele, while dermoid tumors, lipomas, pelvic hamartomas, leiomyosarcoma, and carcinoid tumors have all been documented in literature but are very uncommon. Our case had an epidermoid cyst which is exceptionally uncommon, with a frequency of 4% among patients with Curarrino triad in adulthood. (14)

Currarino traid is rare in adults and should be diagnosed early to prevent life-threatening complications, such as meningitis, rectal fistulas, and malignant transformation. Approximately, 80% of the patients with the classic triad are recognized during the first decade of life. (15) Urinary symptoms and chronic constipation are amongst the most common manifestations of Currarino triad (16) Other clinical presentations include gynecological anomalies, spinal cord tethering, perianal sepsis, and meningitis (17).

Sacral bony defects are pathognomonic for this triad. Pelvic X-ray is the initial imaging modality of choice (18). On MRI, epidermoid cysts appear as a well-defined cystic mass generally with no perilesional edema (19). MRI can also define the extension of presacral mass and may detect other malformations. Prenatal ultrasound and fetal MRI could be used to identify the presence of a presacral mass during in-utero and is indicated when a history of maternal Currarino triad is present. (20)

The variable set of complex anomalies associated with Curranio mandates a multi-disciplinary management approach. Surgical management involves correction of the anorectal malformations before attending to any existing pre-sacral masses (18,21) . The anterior sacral meningocele is surgically managed through the posterior sagittal approach to provide full perineum exposure by incision from the sacrum to the anus. In our case, the trans-dural pathway was accessed by sacral laminectomy, which permits the excision of the intradural epidermoid cyst (18,21). Given the high level of morbidity and mortality associated with this condition, timely diagnosis and precise pre-operative planning are paramount to maximize patient outcomes (22).

Conclusion

Although rare, the possibility of Currarino syndrome should be entertained in adult patients with lower lumbosacral symptoms and a pre-sacral mass. A thorough physical examination and strategic pre-operative planning are mandatory to maximize patient outcomes.

Abbreviations:
CS: Currarino Syndrome;
MRC: Medical Research Council;
MRI: Magnetic resonance imaging;
L2: second lumbar vertebrae.

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