Case Report: Presacral Epidermoid Cyst in a Young Girl with Sacrococcygeal Scoliosis and Anal Sinus

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Abstract

Introduction: Perirectal epidermoid cyst is a rare mass arising from the ectodermal germ cell layer of the hind gut, which is most often found in middle-aged women. It is difficult for us to make an accurate diagnosis and distinguish from other developmental cysts.

Case Presentation: Here, we report a 19-year-old girl who had suffered from sacrococcygeal falls and burns on the left buttocks in the growth experience. There was a scar on the left buttocks, and a soft mass with sacral deformity was seen in the sacrococcygeal region. According to the results of the sacrococcygeal CT and pelvic MRI, the preoperative diagnosis included presacral epidermoid cyst, anal sinus and sacrococcygeal scoliosis. Finally, the cyst was completely resected posteriorly through sacrococcygeal approach. Pathology was verified as an epidermoid cyst. The patient stayed recurrence-free after six months of follow up.

Conclusion: Perirectal epidermoid cyst is a rare mass that should be considered in the differential diagnosis of pararectal cysts. It is of significant for the approach of surgery and prognosis.

Introduction

Epidermoid cyst can be seen in other parts of the body, but the presacral or pararectal epidermoid cysts are extremely rare (1). Presacral cysts or pararectal are described a developmental abnormality in the embryonal phase and considered to originate from caudal embryonic vestiges (2). Generally speaking, presacral or pararectal epidermoid cysts occur mostly in middle-aged women while it is rare presentation in young age group (3). Abnormal masses in the pelvic floor space are often incidentally discovered during routine examination. Most patients with presacral cysts show no clinical symptoms. However, some patients may present with unspecic symptoms resulting from compression of adjacent tissues, including urinary retention, constipation or a palpable mass near anus (4). In order to improve clinical diagnosis strategies and avoid misdiagnosis, we report a rare case of pararectal epidermoid cyst occurring in a young girl.

Case Report

A 19-year-old girl was found to have an abnormal mass in the sacrococcygeal during a health examination, with the main complaint of difficulty in evacuating her bowels for two months. Three months after she was born, the patient suffered from sacrococcygeal deformity due to an accidental fall and had an imaging examination at the local hospital without any therapy. When she was five years old, her left buttock was scalded by boiling water. She has no family history of inflammatory bowel disease or hereditary tumors. No obvious abnormalities were found in the abdominal examination. Her buttocks were obviously asymmetrical, and there was irregular scar hyperplasia and shrinking at the 4 o’clock position, which was about 3 cm away from the anal opening. Physical examination revealed a soft, mobile and poorly circumscribed mass without tenderness, located in the right perirectal region. It was
obviously touched that sacrum and coccyx displacement to the right. Digital rectal examination indicated a bulge in the retro-rectal area, resulting in a mild stenosis of the lumen. (Fig. 1).

Laboratory tests were within normal limits. Colonoscopy showed slight stenosis between the rectum and anal canal without erosion\ulceration or tumors. Three-dimensional computed tomography (CT) scans indicated an abnormal sacral-vertebrae and the sacral canal is partial enlarged and opened. No destruction of local sacral bone was seen (Fig. 2). Magnetic resonance imaging revealed a 55 mm × 40 mm × 35 mm well-circumscribed unicameral cystic mass in the pelvis that adhered to the left rectal wall and posterior to the sacrum, resulting in right-anterior displacement of the rectum. The wall of vagina was compressed to be wrinkled (Fig. 3). There were strips sign with enhancement in enhanced scan extend from levator ani muscle to skin of the left buttock (Fig. 4). Based on the MRI findings, the preoperative diagnosis included presacral epidermoid cyst, anal sinus and sacrococcygeal scoliosis.

Due to the mass had no connection with spine, and the sinus of her left buttock was asymptomatic with no signs of infection. A complete surgical excision thought the posterior trans-sacrococcygeal approach was executed. The patient was positioned in the jackknife position after spinal anesthesia. A longitudinal median incision was made over the mass of body surface localization on the left side of the coccygeal bone. The skin, subcutaneous tissue, fat layer and levator ani muscle were cut layer by layer to the space between sacrum and rectum. The cystic mass was tightly attached to the puborectalis and left posterior side wall of the lower rectum. By utilizing both blunt and sharp dissection to carefully dissect from surrounding structures, the cyst was completely excised from the deep presacral space. The cyst was measured 55 mm × 40 mm × 35mm and filled with bean curd residue like contents material. The rectum was confirmed intact by intraoperative anoscope. The presacral space was stitched in layers with a drainage tube left in situ. On histology, the cystic cavity is covered with squamous epithelium, the contents are gray and white cheese like layered keratinocytes, mixed with exfoliated broken epidermal cells, keratin and cholesterol. The cystic mass was identified as an epidermoid cyst. After operation, the patient was hospitalized for dressing change and bandaging. The patient was discharged 2 weeks after surgery with a drain and stitches removed. Post-surgery for six months, the patient remains recurrence-free.

**Discussion**

The presacral space is a potential triangular space between sacrum posteriorly and the rectum anteriorly, bounded by the peritoneal reflection in the superiorly direction, levator muscles inferiorly (1). Developmental cystic mass arising in the presacral location include a multitude of tumors, including dermoid cysts, epidermoid cysts, chordomas, adrenal rest tumors, anterior sacral meningoceles, cystic hamartomas, tailgut or rectal duplication cysts (5). The epidermoid cyst lie in this space is uncommon with an incidence of 1 in 40000 to 63000 hospital admissions, and 60% perirectal epidermoid cysts are a form of congenital developmental cysts (6).
Perirectal epidermoid cysts are typically slow growing, about 26–50% patients are asymptomatic, most of them were incidentally discovered during imaging (7). Symptomatic compression of pelvic structures by the enlarging cyst may present with urinary complaints, constipation, perianal pain or a palpable mass in the pre-coccygeal region. In case of infection, these masses may result in perianal discharge, fistulous opening and bleeding per rectum. Although epidermoid cysts are common skin lesions, they rarely become squamous cell carcinoma. Malignant tumors arising from epidermoid cysts are reported to appear at a rate of 0.011–2.2% (8, 9). The exact mechanisms by which the epidermoid cysts become malignant tumors remain unclear. However, it may be attributed to the chronic inflammatory responses to repeated cyst ruptures and subtotal resection of the cyst wall (10).

Elevated levels of AFP or HCG may indicate germ cell tumors. Colonoscopy may show extrinsic rectal compression and exclude intestinal space occupying lesions. Transrectal ultrasonography may be useful in assessing the location and extent of small mass and its connection with the anal sphincter (11). CT examination can clearly show the bone destruction of malignant presacral masses. MRI is found to be superior to CT scan in differentiating between any bony, spinal canal or meningeal involvement and its relationship with surrounding soft tissues and organs to determine the surgical plan (12). Although infection, hemorrhage or calcifications in these lesions may alter the signal intensity, subtle changes in signal intensity favor epidermoid cysts. However, these findings are not specific. Except for suspicious cancerous lesions, a preoperative biopsy should not be performed because it can lead to tumor dissemination, abscess, fecal fistula, or meningitis (1).

The choice of surgical path for presacral tumors mainly depends on the location, size and relationship with surrounding tissues and organs. The common surgical approaches are trans sacrococcygeal, trans abdominal, trans sphincter or trans abdominal and trans sacral combination (13). If the mass is small (≤ 10 cm), located at the caudal level (below S4), and does not invade surrounding structures, the sacral approach is usually chosen (14). When the mass is located high (above the sacral level 3), the transabdominal approach is the better option. When the mass is huge or located at the junction, or the boundary with surrounding organs such as ureter and iliac vessels is unclear, and it is difficult to complete the operation through a single approach, the combined approach via the abdominal sacrococcygeal approach can be selected. The sphincter approach is one of options for patients with small, low-lying lesions (15). Only some gynecologists chose transvaginal approach for merits of providing sufficient field of vision, less operative time and less blood loss in low-lying retro-rectal lesion (16).

When presacral epidermoid cyst was complicated with infection and presentation as perianal sinus, it was easily treated as anal fistula or pilonidal sinus (17, 18). When the presacral cyst was closely connected with the coccyx, it was difficult to distinguish from anterior sacral meningocele (19, 20). In female patients, high presacral mass was even misdiagnosed as ovarian cystadenomas (21). The unique feature of this case is that it found a presacral mass with perianal sinus and sacrococcygeal malformation in a young girl. To accurately determine the extent of the mass lesion and to rule out other pathologies, the patient was subjected to several imaging examinations. The MRI indicated a solitary
abscess in the left ischiorectal fossa, which had no sinus interacting with the presacral mass. Given her history of scald, it may be that sterile necrotic tissue accumulated in the perianal sinus. According to the three-dimensional computed tomography scans, an abnormal sacral-vertebrae with the sacral canal partially enlarged and opened, but there was no damage to the sacral surface bone. The wall of cyst was remote from sacral canal, the opened sacral canal was at the S3 level while the cyst was below the S4. The evidence of anterior sacral meningocele was insufficient. Although it is extremely rare, benign cysts can also transform into malignant tumors(22). In our case, a well-defined smooth margin with the absence of invasion may help to differentiate it from malignant masses. This case highlights the importance of pre-operative imaging examination and evaluation to identify the nature of the presacral mass, which is crucial to the way of operation and prognosis.

Nowadays, laparoscopic surgery is known as an option for its minimal invasiveness, reduced risk of complications, and complete tumor removal. Considering the patient was a young girl who had not given birth, in order to avoid damaging the pelvic organ, the surgery was performed by trans-sacrococcygeal approach, which was safe and effective to remove low presacral masses. Because the patient’s coccyx was offset to the right, the incision along the midline of the back did not damage the anococcygeal ligament. However, the cystic lesion was ruptured during the surgery. Cut open the cystic mass to reduce the volume for larger operation space in dealing with giant presacral cyst. In the case of suspected malignant tumors, it should be carefully stripped along the capsule to prevent the tumors from rupturing and spreading. In this regard, the importance of preoperative differential diagnosis is emphasized.

Conclusions

The clinical manifestations of presacral masses are various, and once found, colonoscopy, sacrococcygeal CT and pelvic MRI should be further evaluated. According to the location, size and relationship with surrounding organs, the appropriate surgical path should be selected. It is a significant diagnostic challenge to distinguish an epidermoid cyst from other pararectal cystic mass.

Declarations

Ethical approval

As a case report without Protected Health Information, no ethics approval was required for this project.

Consent For Publication

The patients/participants provided their written informed consent to participate in this study. Written informed consent was obtained from the individual(s) for the publication of any potentially identifiable images included in this article.

Availability Of Data
All datasets generated for this study are included in the article/supplementary material.

Competing Interests

The authors have no conflict of interest, financial or otherwise.

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Author Contributions

QY was the first to treat patients in the clinic, SY and ZJ were involved in managing the patient and assisting in the operation. DW was the main surgeon. ZJ prepared and wrote this report. LL and QL prepared the imaging pictures. DW revised the manuscript as well as acted as the corresponding author. All authors contributed to the article and endorsed the version submitted.

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References


Figures
Figure 1

Pre-operative image of the mass

Figure 2

(A) frontal view and (B) dorsal view: three-dimensional CT scan showed a sacrococcygeal scoliosis below the S2 level (white arrows), the sacral canal is partial enlarged and opened.
Figure 3

Axial (A) T2-weighted imaging there was a well-circumscribed mass compressing on the rectum and displacing it right-anteriorly; Sagittal (B) T2-weighted imaging showed a well-defined mass anteriorly to the sacrum.

Figure 4

Axial T1-weighted pre(A) there was a circular signal (white arrows) on the outside of levator ani muscle with strips signs connecting to skin of the left buttock; Axial (B) contrast enhanced T1-weighted imaging showed enhancement of strips signs.

Supplementary Files

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- CareChecklist.jpg