

A presacral epidermoid cyst presenting with a perineal fistula: report of a case

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Abstract: Presacral epidermoid cysts are rare lesions that become clinically evident only when complicated by pain or infection. Usually they are asymptomatic and diagnosis is unexpected at the time of a gynaecological or ano-rectal examination or as an incidental finding report on a radiological examination such as a pelvic CT scan or MRI performed for other reasons. We report the case of a young male presenting with a perineal fistula. Clinical examination, pelvic MRI and CT scan demonstrated an infected pre-sacral cyst with a anal fistulous tract. Difficulty in localising the cyst, proximity to sacral promontory, the need to preserve the integrity of both the rectal wall and the hypogastric nerves from surgical injury led to adoption of a combined laparoscopic and perineal procedure for its complete excision.

Key words: Presacral epidermal cyst; Retro-rectal space; Laparoscopy; Anal fistula.

INTRODUCTION

The human embryo has a true tail between the 28th and the 35th day of gestation. This is an extension of the primitive hindgut and, being caudal to the subsequent site of development of the anus, it is called the *tailgut*. It normally regresses completely by the 8th week of gestation.¹ A wide range of disembryogenic masses can derive from incomplete regression of the primitive hindgut. Remnants of the tailgut or notochord lead to the development of retro-rectal lesions. Most of them are benign and include epidermal and dermal cysts, rectal duplications, hamartomas and seminal vesicle or Mullerian cysts. A small number of them carry a malignant potential, arising from a benign lesion or from a primitive malignancy, such as a teratoma. These masses are almost exclusively localized in the presacral space which is located behind the posterior side of the rectal fascia, above the Waldeyer fascia, which is 3 cm above the pelvic muscles layer; below the abdominal peritoneal reflexion and in front of the sacrum. The rectal pillars, iliac vessels and ureters surround this space on each side. These masses are rarely found in other locations.²

They can occur at any age but retrorectal cysts are more frequently observed in female patients 12-35 years of age. The male-female ratio is 1:3. Because of their rarity, data on prevalence are lacking in the literature, with an incidence varying from 1.4 to 6.3%.³

CASE REPORT

A young male complaining a perineal fistula was referred to our outpatient department in October 2004. His clinical history included an asymptomatic prolapse of the mitral valve and a malformation of the L3-L4 vertebral bodies. His previous surgical history included appendectomy.

Because of the onset of a perineal fistula associated with sacrococcygeal pain he underwent a careful exploration of the fistulous tract. Subsequent positioning of a drainage tube immediately resulted in the passage of a great quantity of pus and of a hair tuft. Palpation of a retrorectal mass during rectal digital examination lead to the suspicion of a presacral abscess. Therefore the patient underwent a recto-sigmoidoscopy, pelvic CT and MRI scan which showed a presacral fluid mass of 7x2x2 cm without any apparent relationship with the rectum (Fig. 1). The following trans-rectal ultrasound (TRUS) confirmed the presence of a dishomogeneous liquid mass, extending to the pelvic floor without any apparent fistulous communication with the rectal lumen (Fig. 2). Through a posterior parasacral incision a large part of the

presacral mass was removed but the complete excision was not possible as the superior pole resulted too far away. Thus a *Malecot* tube was inserted from the perineal fistula to the cranial remnant of the abscess. Histology performed on the specimen resulted in an *infected epidermoid cyst* without malignant degeneration (Fig 3). Furthermore a following fistulography through the previously positioned tube with contemporary endorectal enema showed an elongated shaped cyst located at S2 level, confirming that no rectal fistula was present (Fig. 4). Eight weeks after the first operation the patient underwent a combined laparoscopic and perineal surgical procedure to completely remove the fistulous tract and the retrorectal cyst. This procedure was performed by an experienced laparoscopist and a colorectal surgeon at the same time. An x-ray of the specimen was done to check its complete excision (Fig. 5). Patient was discharged after 7 days completely recovered.

No signs of infection, fistula, sexual or voiding dysfunction or rectal injury were reported during 4 years of follow up.

DISCUSSION

Presacral cysts are rare entities. Other retro-rectal space lesions include tumors, such as chondromas arising from notochord vestiges and, more frequently in males, anterior

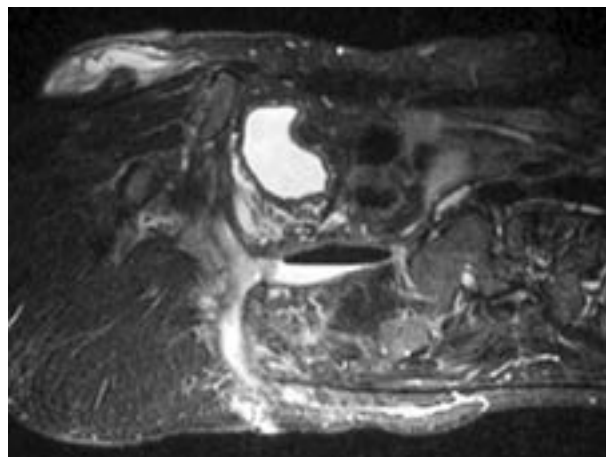


Fig. 1. – With patient in horizontal position, presence of fistulous tract from right gluteal region in communication with elongated vertical cavity containing liquid and air. The image shows no relationship with rectal lumen.

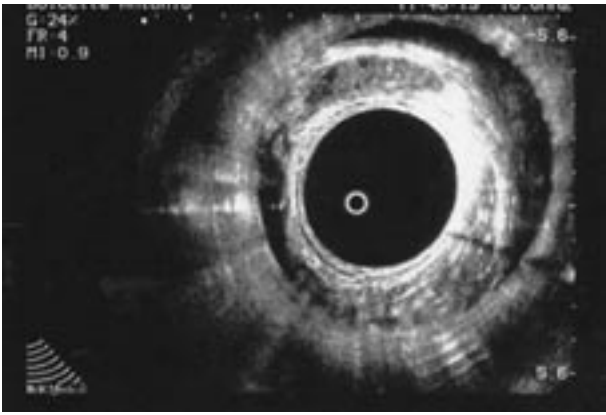


Fig. 2. – With patient in Sims position, evidence of ipoechoic tissue in the posterior perirectal space corresponding to the most caudal portion of the infected cyst. The development of fistulous tract to perineal skin appears to circle the rectum also from the lateral left side.

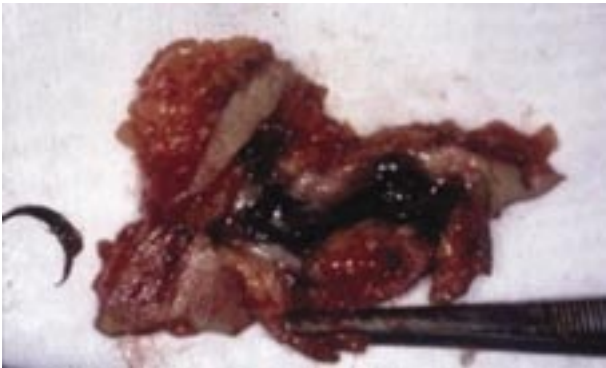


Fig. 3. – Operatory specimen after posterior excision. Notice the presence of hair tuft.

sacral meningocele, neurolemmomas, ganglioneuromas, neurofibrosarcomas, bony cysts or lymph nodes from rectal cancer.³ The exact prevalence is not known but data coming from referral centers estimate an incidence of 1/40000-1/63000 of admitted patients.^{4,5}

Higher incidence in female patients seems to respond to a clinical bias. In fact gynaecological and proctological examination on young girls allows an earlier detection of presacral cysts than in males. Usually asymptomatic, they grow slowly and become clinically evident because of sense of rectal fullness, tenesmus, sacro-coccygeal pain or infection. Although most of presacral lesions present benign features, malignancy can always be possible. Careful rectal examination is essential and represent the first step for the correct diagnosis. Tenderness, mobility and smooth surface represent benign features whereas a hard, sharp, fixed and painful mass suggest the presence of malignancy. As rectal digital examination is sensitive but not specific,⁶ anoscopy and full colonoscopy must be performed in order to exclude a rectal cancer or other colonic lesions. FNAB should be avoided because of high risk of infection (especially if performed through the posterior rectal wall) or malignant seeding. The high incidence of recurrence and the possibility of misdiagnosis⁷ suggest that the complete excision must be the procedure of choice, reserving biopsies of specimen just in cases of great suspicion of malignant tumors that require highly demolitive surgery. Allowing a good vision of the sacral nervous plexus and avoiding large rectal mobilization, the

posterior surgical approach is interesting, but considering our experience it should be reserved for retrorectal cysts whose superior pole is palpable during rectal examination. For lesions not fully reachable at digital exploration, abdominal approach gains a better view of pelvic organs and allows to isolate the cyst completely, once the rectum has been mobilized.

In the case reported the patient was complaining presacral pain, fever and a perineal fistulous orifice. For this reason the patient was firstly treated with the position of



Fig. 4. – Injecting contrast through the perineal fistula (Malecot tube), dullness of fistulous tract for 6 cm. Just right to ano-rectal junction, visualization of vertical shaped cyst whose measures are 7x2x2 cm. The superior pole of the cyst appears 1.5 cm far from upper side of S1 body.

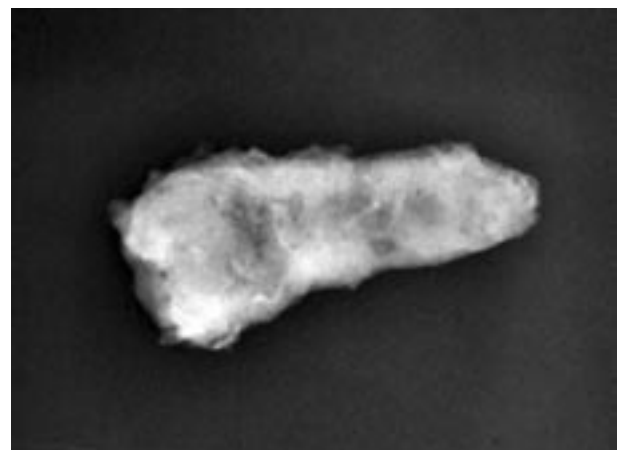


Fig. 5. – Radiologic control after laparoscopic excision of retro-rectal cyst remnant.

a drainage tube. The subsequent output of pus mixed with hair suggested the hypothesis of a disembryogenic infected cyst. TRUS is a sensitive method for analysis of the rectal wall and the perirectal space, helping to distinguish solid from liquid lesions. In our case the position of the cyst, its extension and size and its relationship with the rectal wall were determined. Because of its better definition of the soft tissues, MR allowed to evidence the longitudinal cyst of 7x2x2 cm with an air fluid level and a gross fistulous tract directing to the perineal skin. Because of the vertical development of the cyst with its superior pole at the sacral promontory, an abdominal operation became necessary to remove the cyst remnant combined with a perianal fistulectomy. Laparoscopy allows a good vision of the deepest part of the pelvis preserving nerves, vessels, ureters and providing less discomfort for the patient and a shorter and less expensive hospital stay. Furthermore, rectum mobilization necessary to reach the retrorectal space, results easier and less time-expensive when performed through a laparoscopic dissection. Contemporary anal fistula excision allowed to complete the procedure without necessity of other surgical operations.^{7,8}

For this reasons, when possible, the combination of laparoscopic and pelvic procedure represents the best alternative to classic laparotomy for high presacral infected cysts, avoiding long hospital stay and reducing patient's discomfort.

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Anastomotic-vaginal fistula (AVF) after anterior resection of the rectum for cancer - occurrence and risk factors. *Matthiessen P, Hansson L, Sjødahl R, Rutegård J. Colorectal Dis. EPUB: 2009-02-18.* To assess recto-vaginal fistula after anterior resection of the rectum for cancer with regard to occurrence and risk factors, 20 female patients who developed a symptomatic fistula were compared with 32 who developed conventional symptomatic leakage and 338 who did not leak. Patients with AVF had lower anastomoses and decreased BMI compared with those with conventional leakage. Risk factors for AVF in multivariate analysis were anastomosis < 5 cm above the anal verge, preoperative radiotherapy and UICC cancer stage IV. Previous hysterectomy was not a risk factor. The need for abdominal reoperation and defunctioning stoma is not different from patients with conventional leakage.

9 – BEHAVIOUR, PSYCHOLOGY, SEXOLOGY

Ageing, mate preferences and sexuality: a mini-review. *Oberzaucher E, Grammer K. Gerontology. EPUB: 2009-02-21.* Sexuality never ceases to be part of a relationship. With increasing age, reproduction loses importance, while pair bonding functions remain relevant. The evolutionary constraints that lead to the evolution of sexual reproduction are framed by the better repair mechanisms of fatal mutations, as well as the need for variable immune systems imposed on large organisms by parasites, such as viruses and bacteria. These factors affect mate choice, especially as regards the gene complex that encodes the immune system. The need to increase both the likelihood of gametes to encounter each other as well as sufficient provision of nutrition for the offspring then leads to the evolution of two sexes: large numbers of small mobile sperms ensure that gametes meet, whereas large egg cells full of energy provide for the zygote, thus leading to a developmental advantage. The asymmetric investment in the offspring affects also cognitive strategies. Men place more importance on youthfulness and fertility than women, who regard resource holding potential as a more relevant criterion. Consequently, jealousy is connected in females to endangered access to resources, in males to paternal uncertainty.

10 – MISCELLANEOUS

Endoscopic closure of the natural orifice transluminal endoscopic surgery (NOTES) access site to the peritoneal cavity by means of transmural resorbable sutures: an animal survival study. *von Renteln D, Eickhoff A et al. Endoscopy. EPUB: 2009-02-14.* Endoscopic closure of the transgastric access site is still a critical area of active research and development into NOTES. Endoscopic gastrostomy closure by means of resorbable sutures was performed in 10 female domestic pigs in an animal survival study. Mean suturing time was 26 minutes (range 14 - 35 minutes). One case of gallbladder perforation occurred during peritoneoscopy and the pig was sacrificed due to peritonitis.

Erratum

In Vol 28, issue 2, pag. 50 (Complex pelvic problems - a multidisciplinary perspective), corresponding Author: Marco Soligo Servizio di Uroginecologia, U.O. Ginecologia e Ostetricia, Ospedale San Carlo Borromeo, Milano, *add*
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