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Case report

Surgical management of large, connected perineal and pelvic epidermal inclusion cysts mimicking a dumbbell-shaped lesion in an adult male

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ABSTRACT

Introduction and importance: Epidermal inclusion cysts are a common benign finding, and they are predominantly asymptomatic. They can rarely form in the pelvis or abdomen, however, and may cause symptoms secondary to mass effect. This case highlights management of an anterectal epidermal inclusion cyst connected to the perineal cyst, mimicking a dumbbell-shaped lesion, found in a male.

Case presentation: This is a unique case of a 21-year-old Caucasian male with a palpable perineal mass, lower extremity hypoesthesia, and constipation who was found to have a complex-shaped cyst on computed tomography and magnetic resonance imaging. This was ultimately managed with a two-stage perineal and transabdominal resection.

Clinical discussion: This case highlights that perineal epidermal inclusion cysts may have pelvic extension, especially in patients with additional new-onset neurologic, gastrointestinal, or urologic symptoms. These symptoms should completely resolve after resection. Additionally, resection is recommended to prevent complications including malignant degeneration and fistulization.

Conclusion: This is the first reported case of an anterectal, epidermal inclusion cyst connected to a perineal cyst found in a male. Perineal and pelvic cysts may be synchronous and may be connected through the pudendal canal. These masses can be safely removed via a combined perineal and transabdominal resection. The connecting portion of lesions that have both pelvic and perineal components should be meticulously identified and dissected because even a thin, patent segment – if left unresected – may result in lesion recurrence.

1. Introduction

Pelvic epidermal inclusion cysts are rare ectodermal lesions that may occur without a genetic predisposition [1]. These are often asymptomatic and misdiagnosed due to nonspecific symptoms including urinary, gastrointestinal, or neurologic complaints secondary to mass effect [2,3]. As a result, patients require a thorough diagnostic evaluation and necessitate resection to confirm their etiology. On histology, these lesions characteristically have a thin wall of keratinizing stratified squamous epithelium and contain cream cheese–like keratin material without fluid [4]. In this case, a young, otherwise healthy male patient was found to have large epidermal inclusion cysts in the rectovesical pouch and perineum causing mass effect symptoms. Both cysts were connected by a narrow fibrous band that traversed the pudendal canal. After a two-stage excision of the perineal then the pelvic cysts, the patient's gastrointestinal and neurologic symptoms completely resolved. This case is unique and describes an anterectal, epidermal inclusion cyst connected to the synchronous perineal cyst, mimicking a dumbbell shaped lesion which has not yet been reported. This report is in line with the SCARE criteria [5].

2. Presentation of case

An otherwise healthy 21-year-old male with no past medical or surgical history presented to a university hospital for evaluation of a tender, palpable left buttock mass. He stated that it had been present for several years but rapidly enlarged over the previous six months. He noted recent onset constipation and left lower extremity numbness, exacerbated by sitting. He denied drainage or redness. He denied using any medications and had no pertinent family history of cancers. He denied use of tobacco, alcohol, or recreational drug use. On physical examination, the mass was mildly painful on deep palpation and

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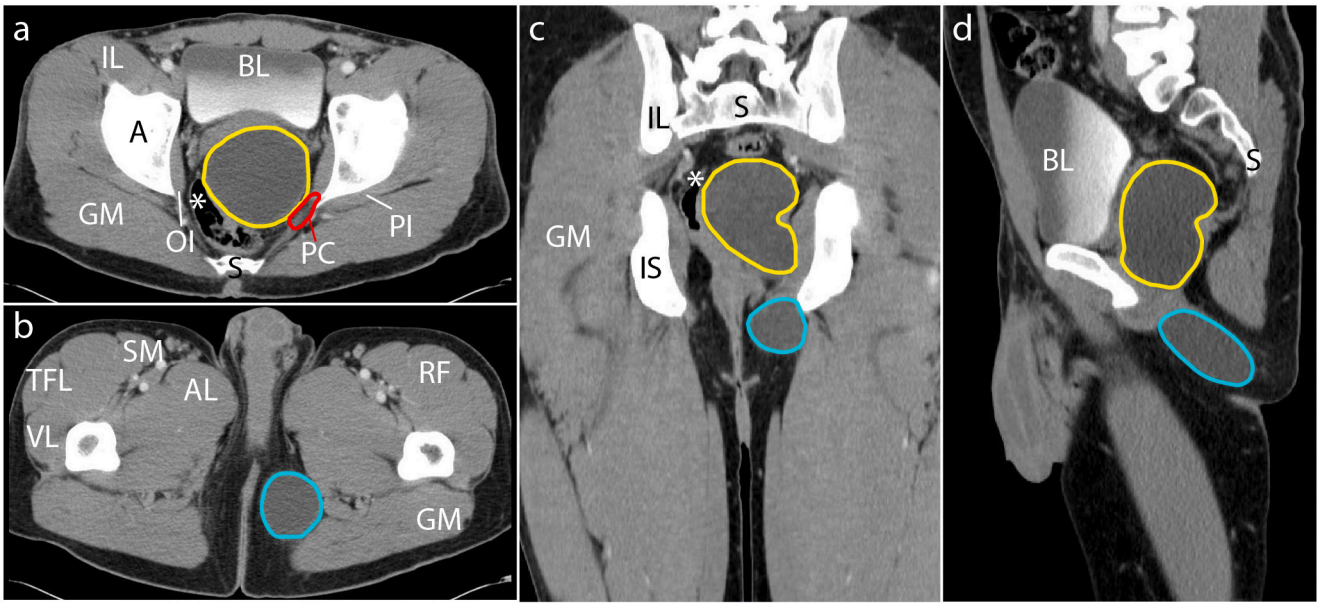

Fig. 1. Preoperative contrast-enhanced CT showing well-defined unilocular hypodense mass with a thin wall anterior to the rectum. a) Axial CT scan of pelvis showing pelvic mass (yellow outline) next to pudendal canal (PC, red outline), bladder (BL), iliacus muscle (IL), acetabulum (A), gluteus maximus muscle (GM), obturator internus muscle (OI), external iliac artery (EIA), piriformis muscle (PI), sacrum (S), and rectum (*). b) Axial CT scan of more caudal section highlighting perineal portion of mass (blue outline), semimembranosus muscle (SM), tensor fasciae latae muscle (TFL), vastus lateralis muscle (VL), adductor longus muscle (AL), rectus femoris muscle (RF), and gluteus maximus muscle (GM). c) Coronal CT scan showing both pelvic (yellow outline) and perineal (blue outline) portions of mass, iliac crest (IL), sacrum (S), ischium (IS), gluteus maximus muscle (GM), and rectum (*). d) Sagittal CT scan showing pelvic (yellow outline) and perineal (blue outline) portions of mass, bladder (BL), and sacrum (S).

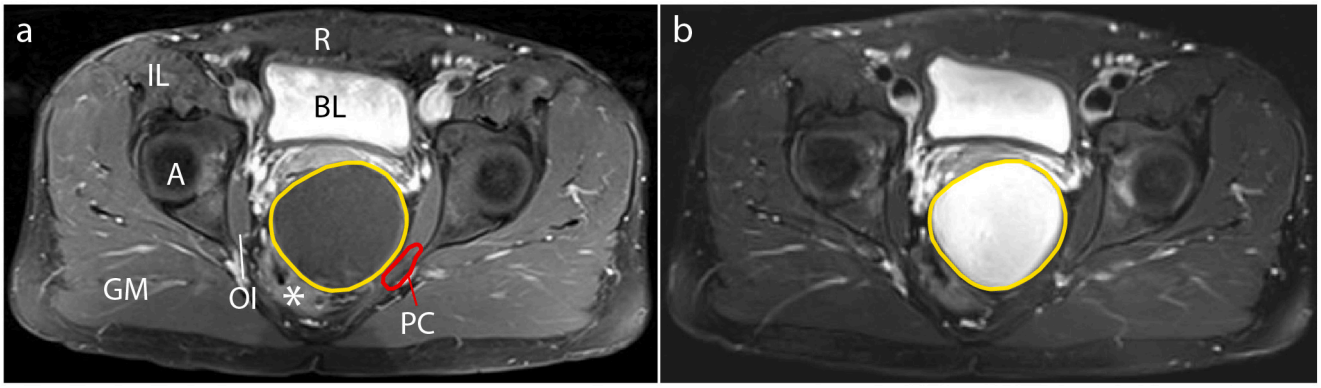

Fig. 2. Preoperative contrast-enhanced T1- and T2-weighted MRI showing pelvic mass. a) Axial T1-weighted MRI showing hypointense homogeneous cystic lesion (yellow outline) without enhancement displacing the rectum (*) and next to the pudendal canal (PC, red outline), rectus abdominis (R), iliacus muscle (IL), gluteus maximus muscle (GM), obturator internus muscle (OI), and acetabulum (A). b) Axial T2-weighted MRI showing homogeneous hyperintense lesion (yellow outline).

measured 4 × 4 cm without overlying skin changes. There were no palpable abdominal masses. There was left lower extremity hypoesthesia in the L5–S4 distribution with preserved motor function exacerbated by sitting, while the right lower extremity examination was unremarkable. No mass or blood was noted on digital rectal examination.

The patient underwent contrast-enhanced computed tomography (CT) which visualized a large, cystic, non-enhancing cystic lesion anterior to the rectum measuring 7.0 × 9.0 cm along with a 4.4 × 4.7 cm cyst in the left perineum (Fig. 1a–d). Contrast-enhanced magnetic resonance imaging (MRI) showed a T1 hypointense, T2 hyperintense homogeneous lesion without enhancement (Fig. 2a–b). The lesions appeared separate from the bladder, urethra, seminal vesicles, prostate, and rectum. Imaging suggested these lesions may have a short neck that penetrated the pelvic floor medial to the inferior pubic ramus (IPR) at the pudendal canal. It was unclear if there was a luminal connection.

No preoperative optimization was warranted. We offered the patient a single-stage perineal and pelvic resection as this was the most efficient approach. The patient preferred that the symptomatic perineal cyst was excised first. The patient elected to delay abdominal incision for social reasons. Given the benign characteristics on imaging, we felt the likelihood of malignancy was low and staging was an acceptable approach assuming frozen section confirmed lack of malignancy.

An incision was made overlying the palpable perineal lesion and dissection was carried out circumferentially around the cyst (Fig. 3a). At the level of the pelvic floor adjacent to the IPR, a discrete, thin, obliterated band was circumferentially dissected and divided under gentle tension (Figs. 3b–4a). Its stump was marked with a permanent stitch and allowed to retract into the pelvis. Intraoperative frozen and final pathology confirmed a benign cyst based on characteristic keratinizing stratified squamous epithelium (Fig. 4b). Postoperatively, the patient experienced moderate improvement in his lower extremity

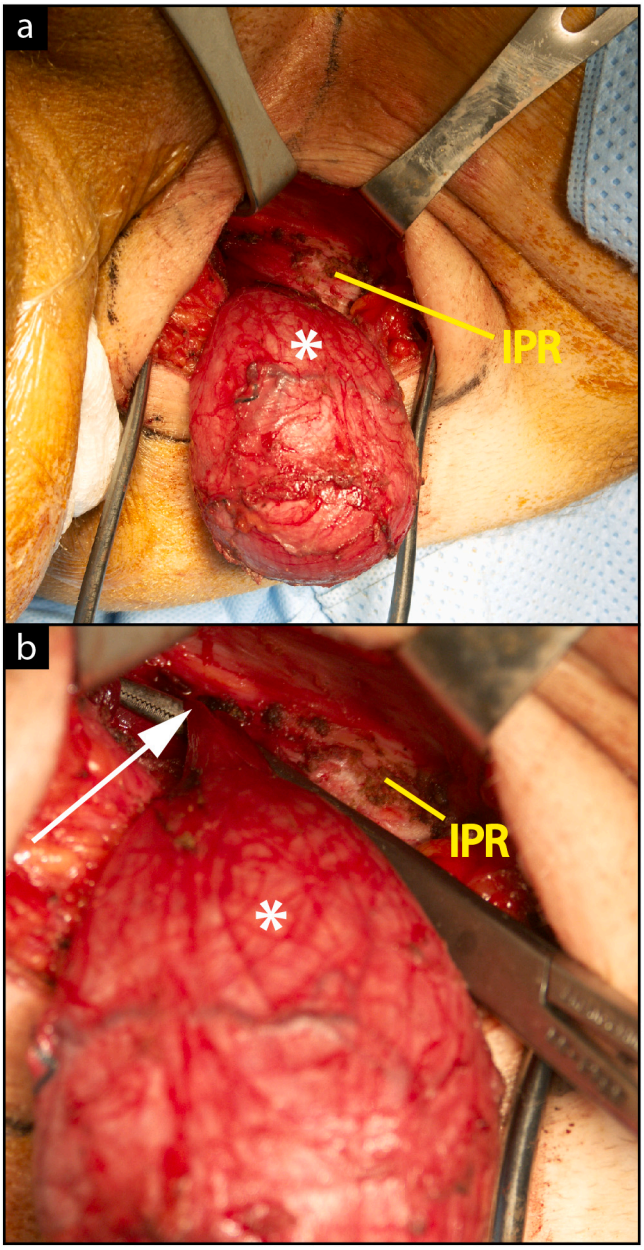

Fig. 3. Intraoperative findings from resection of the perineal portion of the dumbbell-shaped epidermal inclusion cyst. a) The cyst (*) was able to be completely mobilized from the subcutaneous tissue of the left buttock adjacent to the inferior pubic rami (IPR). b) Finding of fibrous band (white arrow) connected to cyst (*) near the inferior pubic rami (IPR).

hypoesthesia.

The second stage was performed six months later. The open approach rather than laparoscopy was elected given the size of the mass and its pressure effect on adjacent pelvic structures. The peritoneum was incised, and a 10 × 7 cm tense pelvic cyst was encountered in the rectovesical pouch filling most of the true pelvis space. Complete circumferential mobilization of the cyst was not possible, so the cyst was incised to allow for decompression. Characteristic white material of cheese-like consistency with numerous fine hairs was evacuated. After cyst content evacuation, the cyst wall was dissected off the pelvic wall and removed, including the entire stump of the connecting band previously marked with a stitch during the perineal operation. Pathology again demonstrated a benign epidermal inclusion cyst without malignant features. No complications or adverse events occurred in either stage of the procedure.

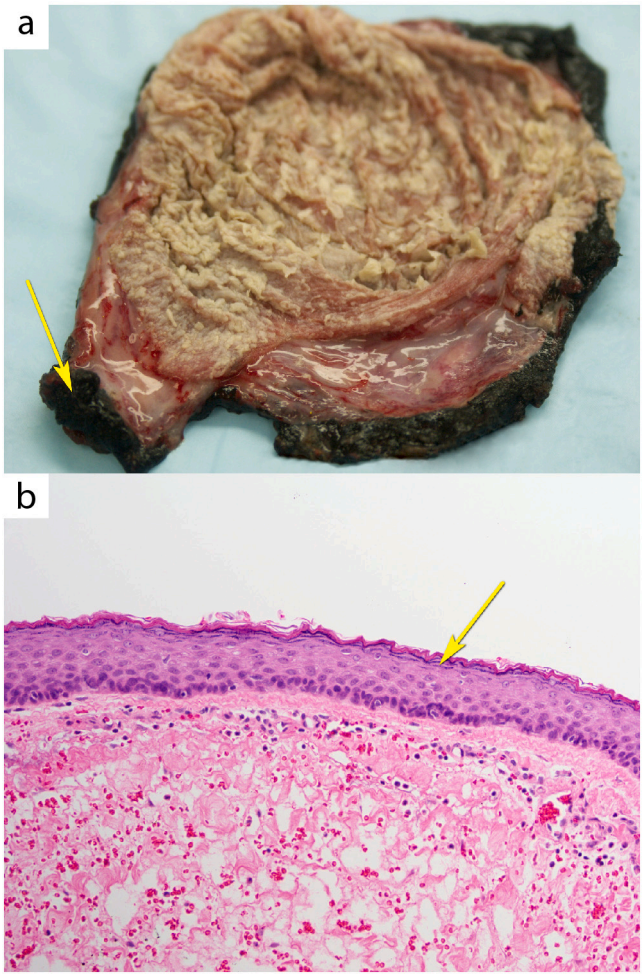

Fig. 4. Additional intraoperative findings and histologic findings from resection of the perineal portion of the dumbbell-shaped epidermal inclusion cyst. a) Perineal cyst containing desquamated material. The obliterated band extending through the pudendal canal is marked with an arrow. b) Microscopy of the cyst wall (200× magnification) with Hematoxylin and Eosin Staining demonstrating keratinizing-stratified squamous epithelial lining (arrow) and a prominent granular layer (image courtesy of Robert A. Robinson, MD).

One month postoperatively, the patient was satisfied as previous neurological and gastrointestinal symptoms had resolved without adverse changes to urinary or sexual function. The patient had follow-up exams with CT imaging annually for 3 years to assure lack of recurrence and was discharged from surgical follow-up. The patient was noted to have no cyst recurrence on a CT performed 8 years postoperatively for other nonspecific abdominal symptoms.

3. Discussion

The incidence of pelvic tumors is not known due to their rarity and often mild-to-absent symptoms. As a result, most literature consists of case reports and series predominantly for retrorectal lesions [2]. Retrorectal masses are estimated to occur 1 in 40,000 hospitalizations, and these lesions are more common in females [6]. There are numerous etiologies that need to be considered in the differential diagnosis. The main classifications include congenital, inflammatory, neurogenic, osseous, and miscellaneous, which includes neoplastic lesions [2]. Epidermal inclusion cysts are considered a subcategory of congenital lesions. In this case, the lesions were found in the rectovesical pouch and perineum. To our knowledge, there have been no previous reports of

cysts connected through the pudendal canal, but there are previous cases of pelvic, dumbbell-shaped lesions and a report of an epidermal inclusion cyst that occurred in a female's retrorectal space and perineum [4]. This is the first case that we encountered in the literature of a male patient with a pelvic epidermal inclusion cyst located anterior to the rectum and connected to a perineal cyst. Based on the location and the size difference of the pelvic and perineal components, we hypothesize that the lesion originated in the perineum. With time, it extended through the pelvic floor along the pudendal canal, mimicking a dumbbell-shaped cyst with subsequent fibrotic obliteration of the connecting neck. Both cystic components were connected by a discrete, obliterated band identified intraoperatively through perineal and abdominal approaches.

Understanding pelvic anatomy, patient presentation, imaging, and histologic findings are important for diagnosis and treatment of these lesions. Digital rectal examination is a necessary component of the physical examination because these pelvic masses can be palpable even in asymptomatic patients [7,8]. Additionally, any mass effect symptoms are often nonspecific; they can include urinary, gastrointestinal, and neurogenic symptoms and may only occur if the cyst compresses surrounding structures or becomes infected. CT or MRI is essential preoperatively to determine the location, and such imaging can assist in differentiating the etiology of the lesion [9,10]. Epidermal inclusion cysts will characteristically appear on CT as a unilocular, hypodense, thin-walled mass filled with fluid density [11]. With contrast-enhanced MRI, these cysts have minimal enhancement of the thin wall and have fluid attenuation, which differentiate them from other lesions such as simple cysts and lipomas. On histology, epidermal inclusion cysts contain characteristic desquamated keratin, cholesterol, water, cellular debris, and a thin wall composed of keratinizing stratified squamous epithelium.

Lesions should not be biopsied if they have typical radiographic characteristics of an epidermal inclusion cyst. Transrectal image-guided biopsy is not recommended due to a high risk of cyst infection. Once infected, pelvic cysts are very difficult to manage surgically. Instead of undergoing biopsy, rare cases of malignant degeneration of an inclusion cyst can be identified on imaging as a focus of irregular wall-thickening on the cyst on imaging [3,12,13].

The accepted treatment of symptomatic lesions or suspected malignant transformation is complete excision of the cyst, via open or a minimally invasive approach, while preserving adjacent viscera, nerves, and vasculature. Damage to surrounding structures and the potential necessity for further operations due to incomplete excision are the two primary risks of cyst removal. Generally, non-infected cysts are easier to dissect and allow for better preservation of the surrounding structures. In young, low-risk patients, cysts should be excised to achieve definitive tissue diagnosis and prevent future complications including fistulization or malignant degeneration [3,12,13]. In high-risk patients, small, asymptomatic inclusion cysts with benign imaging characteristics can be observed. However, if the cyst enlarges or becomes symptomatic, or if the patient prefers to have it removed, resection should be considered.

4. Conclusion

Overall, this case highlights that perineal epidermal inclusion cysts may have a pelvic extension. The definitive treatment is resection. Additionally, this case emphasizes that the connecting region of the lesions that have both pelvic and perennial components should be meticulously identified and dissected, as even a thin patent segment if left unresected, may result in lesion recurrence.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Provenance and peer review

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The study is exempted from ethical approval.

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Evgeny V. Arshava.

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Not applicable.

CRediT authorship contribution statement

Dakota Thompson: Conceptualization, writing, performed literature review.
Neil Wilkinson: Reviewing and editing.
Jennifer Hrabe: Conceptualization, reviewing and editing.
Evgeny V. Arshava: Supervision, conceptualization, reviewing and editing. The surgeon who performed the operation and follow-up.

Declaration of competing interest

The authors have no conflicts of interest to declare.

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References

[1] B. Fakhir, N. Mamouni, N. Bouramdane, et al., A rare case of a giant pelvic retroperitoneal epidermoid cyst, Libyan J. Med. 4 (2) (Jun 2009) 61, https://doi.org/10.4176/090210.
[2] K.G. Hobson, V. Ghaemmaghami, J.P. Roe, J.E. Goodnight, V.P. Khatri, Tumors of the retrorectal space, Dis. Colon Rectum 48 (10) (Oct 2005) 1964–1974, https://doi.org/10.1007/s10350-005-0122-9.
[3] P. Jain, D.K. Pal, Pelvic epidermoid cyst: a rare cause of lower urinary tract symptoms, BMJ Case Rep. 2018 (May 2018), https://doi.org/10.1136/bcr-2017-223258.
[4] C. Palanivelu, M. Rangarajan, R. Senthilkumar, M.V. Madankumar, S. Annapoorni, Laparoscopic and perineal excision of an infected "dumb-bell" shaped retrorectal epidermoid cyst, J. Laparoendosc. Adv. Surg. Tech. A 18 (1) (Feb 2008) 88–92, https://doi.org/10.1089/lap.2007.0010.
[5] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, A. Kerwan, Group S, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (Dec 2020) 226–230, https://doi.org/10.1016/j.ijsu.2020.10.034.
[6] S.W. Jao, R.W. Beart, R.J. Spencer, H.M. Reiman, D.M. Ilstrup, Retrorectal tumors. Mayo Clinic experience, 1960-1979, Dis. Colon Rectum 28 (9) (Sep 1985) 644–652, https://doi.org/10.1007/BF02553440.
[7] B. Böhm, J.W. Milsom, V.W. Fazio, I.C. Lavery, J.M. Church, J.R. Oakley, Our approach to the management of congenital presacral tumors in adults, Int. J. Color. Dis. 8 (3) (Sep 1993) 134–138, https://doi.org/10.1007/BF00341185.
[8] R.J. Spencer, R.J. Jackman, Surgical management of precoccygeal cysts, Surg Gynecol Obstet 115 (Oct 1962) 449–452.

[9] M.T. Loock, P. Fornès, P. Soyer, P. Rousset, L. Azizi, C. Hoeffel, MR imaging features of nongynaecologic cystic lesions of the pelvis, Clin. Imaging 37 (2) (2013) 211–218, https://doi.org/10.1016/j.clinimag.2012.04.023, 2013 Mar-Apr.
[10] R. Al-Shoura, H. Malaekah, W. Al Bassam, Giant retrorectal epidermoid cyst masquerading as a perianal swelling, Case Rep. Surg. 2020 (2020), 5750382, https://doi.org/10.1155/2020/5750382.
[11] D.M. Yang, M.H. Yoon, H.S. Kim, Presacral epidermoid cyst: imaging findings with histopathologic correlation, Abdom. Imaging 26 (1) (2001) 79–82, https://doi.org/10.1007/s002610000118, 2001 Jan-Feb.
[12] D.M. Yang, H.C. Kim, H.L. Lee, S.H. Lee, G.Y. Kim, Squamous cell carcinoma arising from a presacral epidermoid cyst: CT and MR findings, Abdom. Imaging 33 (4) (2008) 498–500, https://doi.org/10.1007/s00261-007-9287-0, 2008 Jul-Aug.
[13] X. Wu, C. Chen, M. Yang, X. Yuan, H. Chen, L. Yin, Squamous cell carcinoma malignantly transformed from frequent recurrence of a presacral epidermoid cyst: report of a case, Front. Oncol. 10 (2020) 458, https://doi.org/10.3389/fonc.2020.00458.