



Neuroendocrine tumour developing within a long-standing tailgut cyst: case report and review of the literature

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Abstract

A tailgut cyst is a rare congenital lesion that can develop in the presacral space from the remnants of an embryonic hindgut. It is unusual for malignant change to occur in a tailgut cyst. We report a case of a large long-standing tailgut cyst, which was removed during a laparotomy. Histopathology showed a well-differentiated neuroendocrine tumour (primary carcinoid tumour) arising in a tailgut cyst. We reviewed the English literature for all adult cases with this condition. All original articles were reviewed, and data were compiled and tabulated. Including this report, 29 cases of NET developing in a tailgut cyst were found in the English literature. Tailgut cysts have been reported as more common in females, with a mean age of presentation in the fifth decade (Devine, in: Zbar A, Wexner S (eds) *Coloproctology*. Springer specialist surgery series, Springer, London, 2010; Hjermstad and Helwig in *Am J Clin Pathol* 89:139–147, 1988). Tailgut cysts may undergo malignant change including adenocarcinoma, sarcoma, and NET (Mathis et al. *Br J Surg* 97:575–579, 2010; Messick in *Dis Colon Rectum* 61:151–153, 2018; Patsouras et al. in *Colorectal Dis* 17:724–729, 2015; Chereau et al in *Colorectal Dis* 15:e476–e482, 2013). It is difficult to estimate the true incidence of malignant change in a tailgut cyst, with the literature reports only limited to case reports and small-case series. Although rare, our case confirms need to consider the possibility of a malignant component, even in a benign process such as a tailgut cyst. This prompts consideration for upfront definitive management.

Keywords Neuroendocrine · Tailgut cyst · Presacral cyst

Introduction

A tailgut cyst is a rare benign congenital lesion that can develop in the presacral space from the persistent remnants of an embryonic hindgut [2, 7]. A tail exists in the human embryo at the gestational age of 28–35 days. It has been hypothesised that the development of a tailgut cyst is due to an incomplete regression of the tail extension of the hindgut by about the eighth gestational week. Most of the literature has described the presence of tailgut cysts mainly in middle-aged women [2, 7, 8]. The presacral space is a potential space delineated by the rectum anteriorly, the presacral fascia (Waldeyer fascia) and sacrum posteriorly, the peritoneal

reflection superiorly, the perineal muscles inferiorly, and the iliac vessels and ureters laterally [2, 7, 9, 10]. Other retrorectal tumours include teratomas, duplication, neuroenteric, dermoid, and epidermoid cysts.

Lesions in the presacral space can be cystic or solid, and can cause symptoms such as abdominal pain, urinary symptoms, menstrual issues, constipation, and anorectal discomfort. There are at least 25 different histologic types of presacral lesions reported, and over half carry malignant diagnoses [11]. Congenital tailgut cysts are the most frequent presacral tumour type. It is rare for malignant transformation (such as neuroendocrine tumour (NET), carcinoma, or adenocarcinoma) to occur in the presacral space; they are most often associated with tailgut cysts when they occur here [12]. It is thought that NET developing in the presacral space could be secondary to direct extension or metastatic invasion of a primary rectal malignancy [13].

We present a rare case of NET within a tailgut cyst without evidence of a primary tumour source or metastatic disease. It was also unique that the cyst has been long-standing for many years, thus surmising the possibility of a rare

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malignant change from an otherwise benign process. We also reviewed the English literature for all reported cases of tailgut cyst with particular attention to any incidental NET on histopathology.

Methods

A review of the literature a MEDLINE search for cases of NET arising in a tailgut cyst was undertaken. All original articles were reviewed, and data were compiled and tabulated qualitatively. The keywords search comprised of “neuroendocrine” or “carcinoid” tumour, and “retrorectal”, “tailgut” or “presacral” (and related linkage terms) cyst. The articles were reviewed to only include publications that reported cases of NET arising in a tailgut cyst. Cases for inclusion were those in the English language and involving patients aged 19 years and older. Patient age, gender, evidence of sacral/coccyx invasion, pre-operative biopsy, management via surgical resection, grade of tumour, Ki67 index, evidence of metastases (pre- and post-operative), follow-up time, and management were recorded where available. NET grading was scored using the WHO classification system [14].

Review of the literature

Including this report, 29 cases of NET developing in a tailgut cyst were found in the English literature. The characteristics of the cases are tabulated in Table 1. Two papers [3, 5] did not provide any information aside from the age range and/or gender of the patient, and thus were not included.

There was a greater proportion of females (17/27; 63%) having NET developing within tailgut cysts when compared to males (10/27; 37%). The average age of females was 40.8 years (range 19–73) and 54.6 years for males (range 35–68). The diagnostic imaging modality most utilised for visualisation of the presacral cyst was computed tomography (CT), which was used in 15 of 23 cases (65%) with information available; however, it was not always the first investigation depending on the clinical situation. There were various presenting symptoms related to the development of a tailgut cyst, including pain (abdomen, pelvic, hip, back, and anal), bowel changes (diarrhoea and constipation), urinary tract issues (recurrent infections and retention), and lower limb neurological symptoms. Two (2/27; 7%) patients were diagnosed with a presacral cyst in an acute setting. Three (3/27; 11%) patients were known to have metastases prior to surgical resection of the tailgut cyst. Ten (10/27; 37%) patients had sacral or coccyx involvement. Three (3/27; 11%) patients had pre-operative metastases. Eight (8/27;

30%) patients underwent a pre-operative biopsy, which was diagnostic of NET.

All patients except one underwent surgery; one received pre-operative treatment (chemotherapy) prior to surgical resection. NET grade from Ki-67 proliferation indexing was reported in 16 cases (all in publications after 2005); there were eight (8/16; 50%) low- and eight (8/16; 50%) intermediate-grade NET on histology. There was no standard approach for the presacral excision, with various methods described or it was not detailed. It was difficult to determine the largest NET resected as not all reports specified the size of NET from histological examination versus the size of the excised cyst. Five (5/27; 19%) patients had post-operative treatment including chemotherapy and/or a somatostatin analogue. Mean follow-up time was 23.7 months (range 3–79 months). Two (2/27; 7%) patients had post-operative recurrence of the NET. Six (6/27; 22%) patients were found to have metastases not seen pre-operatively.

Case report

A 33-year-old woman originally from Afghanistan initially presented with a history of intermittent lower abdominal pain and dysmenorrhoea. Previous pelvic ultrasounds had described polycystic ovaries but no abnormal pelvic masses. The patient had no significant past medical history and there was no known family history of malignancy. A further pelvic ultrasound then noted a large solid and avascular mass in the left adnexa. A CT scan confirmed a large hypodense mass of maximal diameter 11.1 cm within the presacral space that appeared extra-ovarian in origin, closely abutting the right lateral rectal wall, and posterior to the uterus and vagina. She underwent a diagnostic laparoscopy by the gynaecology unit, which demonstrated a firm 10 cm retroperitoneal presacral mass separate from the uterus and bilateral ovaries, with apparent displacement of the rectum. No biopsies were taken at this time. Given its location and relationships, the mass was deemed as not gynaecological in origin. She was referred to another unit for consideration of surgical management, but declined intervention at that time. Serial imaging (via ultrasound and CT scanning) was otherwise performed, with lack of interval growth and a stable septated appearance, with scalloping of the adjacent sacrum suggesting a chronic process.

Eight years after her initial presentation, she was referred to our Colorectal unit with a recurrence of her central abdominal pain, which causing more discomfort on the right side compared to the left. She had no symptoms of obstruction, but described occasional dysuria. MRI characterised the mass as a complex cystic mass containing an internal foci of fat signal measuring 1.8 cm in diameter anteroinferiorly (Fig. 1a–c). The posterior margin

Table 1 Previous cases of NET developing in a tailgut cyst

References	Years	Age	Sex	Presenting symptoms	Past medical history	Family history	Imaging	Sacral or coccyx invasion	Pre-operative metastases	Tumour markers	Pre-operative biopsy	Surgical resection
Schnee et al. [19]	1994	61	M	Sacral mass detected during workup for prostate disease	Neurogenic bladder and hydrourephrosis; transurethral prostate resection (benign); partial gastrectomy for peptic ulcer disease	NA	Bone scan (for suspected sacral metastasis); CT; MRI; angiography (with pre-operative embolisation)	Yes	No	NA	No	Yes
Horenstein et al. [20]	1998	19	F	Pelvic pain, irregular menstrual cycles	NA	Presacral masses; sister with meningocoele	U/S	No	No	NA	No	Yes
Prasad et al. [7]	21	F		Pelvic pain	NA	NA	U/S	No	No	NA	No	Yes
	69	F		Painful bowel movements and mild rectal bleeding (12 months)	NA	NA	CT	No	No	NA	NA	Yes
Oyama et al. [12]	2000	52	M	Diarrhoea, numbness and pain extending down the left leg, and a prominent tender bulge on the left buttock (3 years)	NA	NA	CT	Yes	NA	NA	Yes	Yes
Mourra et al. [21]	2003	68	M	Anal pain (3 years), increasing over 1 year and associated with mucous discharge	Haemorrhoidectomy (20 years prior)	NA	CT	Yes	No	NA	NA	Yes
Jacob et al. [22]	2004	42	F	NA	NA	NA	NA	Yes	No	NA	NA	Yes
Song et al. [23]	2004	41	F	Acute presentation—perianal pain	Nil	NA	None—sigmoidoscopy only	No	No	Normal	No	Yes
Luong et al. [24]	2005	37	M	Increasingly severe back pain with left leg irradiation (2 years)	Nil	NA	CT; MRI	Yes	Yes—liver, lung, nodes, muscle, bone	Elevated serum chromogranin A	Yes—of presacral lesion and liver metastasis	No
Mathieu et al. [25]	2005	49	F	Rectal fullness and irregular mucus emission (few months)	Nil	NA	CT; MRI; 18F-FDG PET	No	No	Normal	No	Yes
Liang et al. [26]	2008	51	F	Left hip pain (3 years)	Peripheral vascular disease; hysterectomy for uterine leiomyoma	No	CT; MRI	No	No	NA	Yes	Yes

Table 1 (continued)

References	Years	Age	Sex	Presenting symptoms	Past medical history	Family history	Imaging	Sacral or coccyx invasion	Pre-operative metastases	Tumour markers	Pre-operative biopsy	Surgical resection
Stefano et al. [27]	2010	73	F	Worsening back and pelvic pain (3 years)	Ulcerative colitis; essential thrombocythaemia; renal Ca; stapled haemorrhoidectomy	NA	CT; MRI; flexible colonoscopy with EUS; OctreoScan scintigraphy	No	No	Elevated neuron-specific enolase	Yes	Yes
Spada et al. [28]	2011	63	F	Increased lumbar pain; peri-umbilical mass	Hysterectomy for uterine fibromas at 45 years of age; slipped lumbar disc	None	MRI; U/S	No	No	Elevated CA19-9	No	Yes
Niazi et al. [29]	2011	28	F	Persistent rectal pain during evacuation	Birth to twins	NA	CT; ¹¹¹ In-Octreotide scintigraphy	No	Yes—liver	Normal	No	Yes
				Worsening back pain, gait difficulties, recurrent urinary tract infections, bowel dysfunction	None	None	XR (spine); CT; MRI	Yes	No	NA	Yes	Yes
Damato et al. [30]	2013	24	F	NA	NA	NA	NA	Yes	No	NA	NA	Yes
Charalampakis et al. [31]	2014	35	M	Acute presentation—urinary retention	None	NA	US; MRI	No	No	Normal	No	Yes
Kim et al. [32]	2014	49	M	Constipation (few months)	Nil	NA	EUS; MRI	No	No	Normal	No	Yes
Abutkar et al. [33]	2014	61	M	Lower back pain, rectal pain, increased defecation urgency (3 weeks)	NA	NA	Barium enema; MRI; CT; NM MIBG (to rule out metastases)	No	No	NA	Yes	Yes
Mitsuyama et al. [34]	2015	53	M	Left gluteal pain, impaired defecation urge, perianal and gluteal sensory disturbance (1 year)	None	None	CT; MRI; PET	Yes	No	Normal	Yes	Yes
Acar et al. [35]	2017	39	F	NA	NA	NA	⁶⁸ Ga-DOTATATE PET/CT (for metastases)	NA	NA	NA	NA	Yes
Mora-Guzmán et al. [36]	2017	56	F	Anal discomfort, supuration (2 months)	? Anal fistula	NA	EUS; MRI	No	No	Normal	No	Yes
Al Khaldi et al. [37]	2018	53	M	Low back pain (3 years)	Congenital renal malformation (operated on)	Prostate Ca	CT; MRI	Yes	No	NA	No	Yes
Iwata et al. [8]	2018	25	F	Acute presentation—coccydynia, urinary frequency post fall; worsening pain	None	NA	XR pelvis; CT; MRI; ¹⁸ F-FDG PET/CT	No	No	Normal	Yes	Yes

Table 1 (continued)

References	Years	Age	Sex	Presenting symptoms	Past medical history	Family history	Imaging	Sacral or coccyx invasion	Pre-operative metastases	Tumour markers	Pre-operative biopsy	Surgical resection	
Erdrich et al. [38]	2018	77	M	Left hip pain radiating to knee	Squamous cell carcinoma, prostate Ca, melanoma	Glioblastoma multiforme (son), prostate Ca (father), lung Ca (maternal aunt), ovarian Ca (maternal grandmother), colon Ca (maternal grandfather)	CT; MRI; Ga68 DOTATATE PET/CT	No	Yes—liver	Normal	Yes	Yes (resection tailgut cyst and liver metastases); cholecystectomy due to position of liver metastases	
Present case	2018	33	F	Abdominal pain, dysmenorrhoea	None	None	U/S; CT; MRI	No	No	Normal	No	Yes	
References	Years	Operation	Size of resected specimen	Time of symptoms to resection	Clear margins on resection	NET grade	Ki-67 of tailgut cyst NET	Immediate post-operative management	Follow-up term	Recurrence	Post-operative metastases	Further management	Other notes
Schnee et al. [19]	1994	Transperitoneal and posterior en bloc resection of mass (adherent to rectum and thus requiring abdominal noperineal resection); S2-24	18 × 14 × 6 cm (amputated sacrum, infiltrating lobular tumour mass, attached bowel loop, adherent skeletal muscle, soft tissue)	NA	NA	NA	NA	Surveillance	24 months	No	No	Nil	
Horenstein et al. [20]	1998	Resection of lesion	NA	NA	NA	NA	NA	Nil	48 months	No	No	NA	Lesion was initially surgically explored and biopsies (without resection). The patient then underwent two cycles of induction chemotherapy (cisplatin, VP-16, ifosfamide) with no effect; an additional three cycles of adriamycin, dacarbazine, cytoxan also had no effect. The patient then underwent surgical excision
		Resection of lesion	NA	> 1 year	NA	NA	NA	Nil	36 months	No	No	NA	

Table 1 (continued)

References	Years	Operation	Size of resected specimen	Time of symptoms to resection	Clear margins on resection	NET grade	Ki-67 of tailgut cyst NET	Immediate post-operative management	Follow-up term	Recurrence	Post-operative metastases	Further management	Other notes
		Resection of lesion	NA	NA	NA	NA	NA	Nil	13 months	Yes	Breast	NA	Breast lumps noted 12 months post with similar morphology to the presacral tumour; at 13 months post, CT showed recurrence of presacral tumour
Prasad et al. [7]	2000	Resection of lesion	1.5 cm (NET)	> 1 year	Unclear—second procedure required	NA	NA	NA	24 months	No	No	NA	
Oyama et al. [12]	2000	NA	NA	> 3 years	NA	NA	NA	Surveillance	6 months	NA	NA	NA	At time of follow-up, diarrhoea symptoms resolved, however numbness and pain extending down the left leg continued
Mourra et al. [21]	2003	Posterior resection of retrorectal lesion and excision of S5	1.2 cm firm nodule within a 2 × 2 cm piece of fibrous tissue with S5 attached	> 3 years	Yes	NA	NA	Surveillance	12 months	No	No	Nil	
Jacob et al. [22]	2004	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	NA	
Song et al. [23]	2004	Abdomino-perineal resection	NA	Acute presentation	NA	NA	NA	Surveillance	> 15 months	No	Liver, brain	Liver metastasis (detected 1-year post-operatively); chemotherapy with 5-fluorouracil, 3 months post chemotherapy, developed diplopia—brain metastasis detected—palliative radiation	

Table 1 (continued)

References	Years	Operation	Size of resected specimen	Time of symptoms to resection	Clear margins on resection	NET grade	Ki-67 of tailgut cyst NET	Immediate post-operative management	Follow-up term	Recurrence	Post-operative metastases	Further management	Other notes
Luong et al. [24]	2005	–	Not resected	>2 years	–	1	2.9%	–	–	–	–	–	Not operated. Post biopsy was treated with long-acting somatostatin analogue. Represented 15 months post initial presentation with deterioration in clinical condition and diffuse pain. Increased node size and chromogranin A level. Treated with somatostatin analogue but then declined treatment after 3 months
Mathieu et al. [25]	2005	Resection of lesion	Cystic lesion with diameter 2.5 cm containing 3 mm carcinoid tumour	NA	Yes	NA	NA	NA	24 months	No	No	NA	
Liang et al. [26]	2008	Laparotomy for resection of mass	4.6 × 4.1 × 3.9 cm (soft tissue + 20% cystic space)	>3 years	Yes	1	<1%	NA	NA	NA	NA	NA	
Stefano et al. [27]	2010	Laparoscopic resection of presacral mass	3.7 × 3 × 2.5 cm (mass)	>42 months	NA	1	<2%	Surveillance	5 months	No	No	Nil	
Spada et al. [28]	2011	Bilateral adnexectomy	5.8 × 3.5 × 4.3 cm (cystic/solid mass); 1.5 cm (NET)	NA	Yes	1	<2%	Surveillance	25 months	No	No	Nil	
		Posterior approach with intersphincteric and para-sacro-coycegeal excisions	NA	>1 month	NA	2	18%	Chemotherapy (carboplatin, etoposide)	79 months	Yes	Left adnexa, multiple bone metastases, nodes	Peptide receptor radionuclide therapy; hysterectomy, bilateral adnexectomy; long-acting somatostatin analogue therapy	
Niazi et al. [29]	2011	T10-S4 laminectomy and duraplasty; dural repair patch	NA	NA	NA	1	Low grade	Physiotherapy, surveillance	12 months	No	No	Nil	
Damato et al. [30]	2013	Radical excision	NA	NA	NA	NA	NA	Surveillance	3 months	No	No	Nil	

Table 1 (continued)

References	Years	Operation	Size of resected specimen	Time of symptoms to resection	Clear margins on resection	NET grade	Ki-67 of tailgut cyst NET	Immediate post-operative management	Follow-up term	Recurrence	Post-operative metastases	Further management	Other notes
Charalampakis et al. [31]	2014	Laparotomy	10×8.5×1.1 cm (collapsed cyst) with maximal 6 mm thick wall	Acute presentation	NA	1	Low grade	Surveillance	36 months	No	No	Nil	
Kim et al. [32]	2014	Transanal excision	NA	NA	NA	2	4%	NA	24 months	See note	NA	NA	One month after surgery, a residual mass in the rectum was noted and monitored with serial imaging as the patient refused additional surgery (unclear if NET or not). No progression was otherwise noted in the 2-year follow-up period
Abukar et al. [33]	2014	Abdominoperineal resection	5.9×4.2×2.7 cm (tumour mass)	>3 weeks	No	1	Low grade	NA	NA	NA	NA	NA	
Mitsuyama et al. [34]	2015	Osteoplastic sacral laminectomy	NA	>1 year	NA	2	12.5%	Complicated by bladder dysfunction, CSF leak; re-operation completed and started somatostatin analogue treatment	28 months	No	Lymph nodes		
Acar et al. [35]	2017	NA	NA	NA	NA	NA	2–5%	Chemotherapy	48 months	NA	Breast, retro-orbital, cardiac, skin, subcutaneous	NA	
Mora-Guzmán et al. [36]	2017	Posterior approach; partial coccygectomy	4.5×2.5 cm (solid lesion)	>2 months	NA	1	<2%	NA	7 months	No	No	NA	
Al Khaldi et al. [37]	2018	Coccygectomy; <i>en bloc</i> posterior surgical resection of mass	7.6×3.9×2.5 cm (specimen), coccyx 3.7×1.0×2.7 cm	>6 months–3 years	Yes	2–3	5–10%	Surveillance	>24 months	No	Lymph nodes (left groin), lungs, liver	Somatostatin analogue; chemotherapy, PPRT due to progressive disease	

Table 1 (continued)

References	Years	Operation	Size of resected specimen	Time of symptoms to resection	Clear margins on resection	NET grade	Ki-67 of tailgut cyst NET	Immediate post-operative management	Follow-up term	Recurrence	Post-operative metastases	Further management	Other notes
Iwata et al. [8]	2018	Posterior approach	6.7 × 2.0 × 1.8 cm (specimen)	> 8 months	NA	2	8%	NA	NA	No	NA	NA	
Erdrich et al. [38]	2018	Laparotomy	NA	NA	NA	2	8.6%	NA	NA	NA	NA	NA	Liver metastases taken at time of tailgut cyst resection had Ki-67 of 6.4% (grade 2 NET)
Present case	2018	Laparotomy	12.8 × 8.5 × up to 2.1 cm (cystic structure); 1.0 × 1.5 (NET)	> 8 years	Yes	1	1–2%	Surveillance	7 months	No	No	Nil	

NET neuroendocrine tumour, CSF cerebrospinal fluid, NA not available, M male, F female, CT computed tomography, MRI magnetic resonance imaging, U/S ultrasound, XRX-ray, EUS endoscopic ultrasound, PET positron emission tomography, NM MIBG nuclear medicine metatodobenzylguanidine, 18F-FDG fluorodeoxyglucose, FNA fine-needle aspiration

of the lesion abutted the sacrum and coccyx; there was no communication with the central spinal canal. The cyst displaced the uterus anteriorly, displaced, and compressed the bladder anteriorly, and compressed the rectum at the left anterolateral margin of the cyst without evidence of upstream bowel dilatation.

Given her ongoing and worsening symptoms, the patient subsequently opted for surgical resection. A midline laparotomy was performed considering the size of the lesion. There were omental adhesions to the large cyst and careful dissection performed to gain access to the pelvis. Due to its size, we decompressed the cyst through a small trephine, suctioned the clear fluid content, and closed the defect with 3–0 PDS to control any leakage. The mass itself was easily separated from the rectum and pelvic sidewalls. Due to the size of the dissection bed, a Jackson-Pratt drain placed left to the posterior rectum and into the pelvis. On digital rectal examination, there was a right posterolateral fistula opening in the anal sphincter, but no tracts noted. No intervention was performed on this at the time of operation.

Macroscopically, the collapsed cyst structure was 12.8 × 8.5 × 2.1 cm in size, was multiloculated with a wall thickness ranging from 2 to 15 mm, and had both solid and cystic components. Histologically, the cyst was surrounded by fibrofatty tissue and skeletal muscle. It was mostly lined by non-neoplastic squamous epithelium showing surface keratosis and hyperplasia and had focal lining of some locules by cytologically bland-ciliated cuboidal cells with scant goblet cells (Fig. 2a). There was one focus within the adjacent fibrous wall, which showed an infiltrative neoplasm (Fig. 2b, c). This was described as solid nests, “ribbon like” cords as well as some tubular structures of mildly pleomorphic cuboidal to columnar cells. Focally, the cells had more abundant eosinophilic cytoplasm. There were less than 1 mitoses per 10 high-power fields. There was no necrosis. The lesional cells surrounded a nerve fibre, but did not appear to show perineural invasion. The focus of tumour was approximately 1.0 × 1.5 cm (microscopically) with no lymphovascular space invasion. The neoplastic cells had diffuse strong staining with cytokeratin AE1/3 (Fig. 2d) and neuroendocrine markers’ chromogranin (Fig. 2e) and synaptophysin (Fig. 2f). There was diffuse weak staining with CDX2 and ER. There was patchy (< 10%) staining with CK7 and the cells were negative with CK20. The Ki-67 proliferating index was noted to be 1–2%. The neoplasm was described as a well-differentiated NET arising in the vicinity of a benign cyst. The NET was approximately 2 mm clear of the apparent surgical resection margins.

Tumour marker values were unremarkable. Pre-operative staging CT scans did not locate any evidence of malignancy throughout the entire time period. Gastroscopy and colonoscopy were unable to identify a mucosal source for the NET. Given these findings and with the resected sample clear of

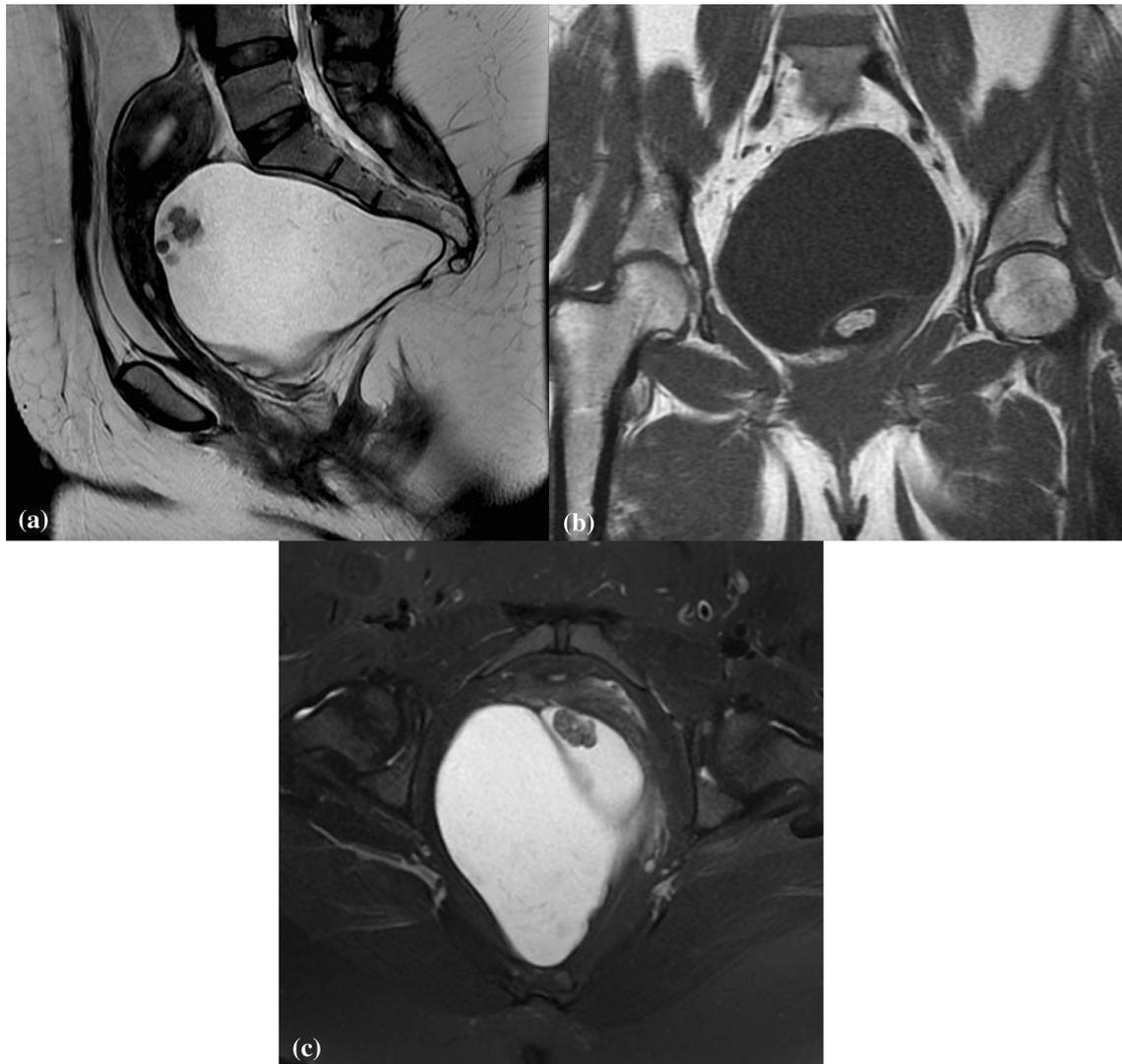


Fig. 1 Magnetic resonance imaging obtained 3 months prior to operative resection measured a persistent and septated cystic mass in the pelvis as $11 \times 11 \times 12.6$ cm. **a** T2-weighted sagittal image of the mass shows the posterior margin of the lesion abutting the sacrum and coccyx with acute angulation of the sacrococcygeal junction. **b** Coronal

image of the mass showed a T1 hyperintense mildly lobulated focus anteroinferiorly measuring 1.8×1.1 cm, demonstrating fat saturation. **c** Axial T2 fat saturated image shows the fat focus in the area, where it was seen previously on CT imaging from 3 years prior to operative resection

neoplastic cells in its margins, the patient was surveyed with interval CT scan.

Discussion

A tailgut cyst is an example of a retrorectal tumour that forms in the presacral space and is derived from the incomplete regression of the embryonic hindgut [2, 11, 12]. Overall, tailgut cysts have been reported as more common in females, with a mean age of presentation in the 5th decade [1, 2]. Tailgut cysts may undergo malignant change including adenocarcinoma, sarcoma, and NET [3–6]. It is

difficult to estimate the true incidence of tailgut cysts having malignant transformation, with the literature containing mostly single-case reports and small-case series that include patients with NET. Most NET occurring in the presacral space are associated with local extension of a primary rectal carcinoid or metastasis from an alternative source [13]. Thus, primary NET of the presacral space is rare.

A few retrospective reviews have attempted to review the incidence of malignant change in tailgut cysts overall. Mathis et al. [3] reported 31 patients (28 women) with tailgut cysts over a 23-year period and across three institutions. All patients underwent surgical resection, and four of them (13%) had developed malignant change: adenocarcinoma

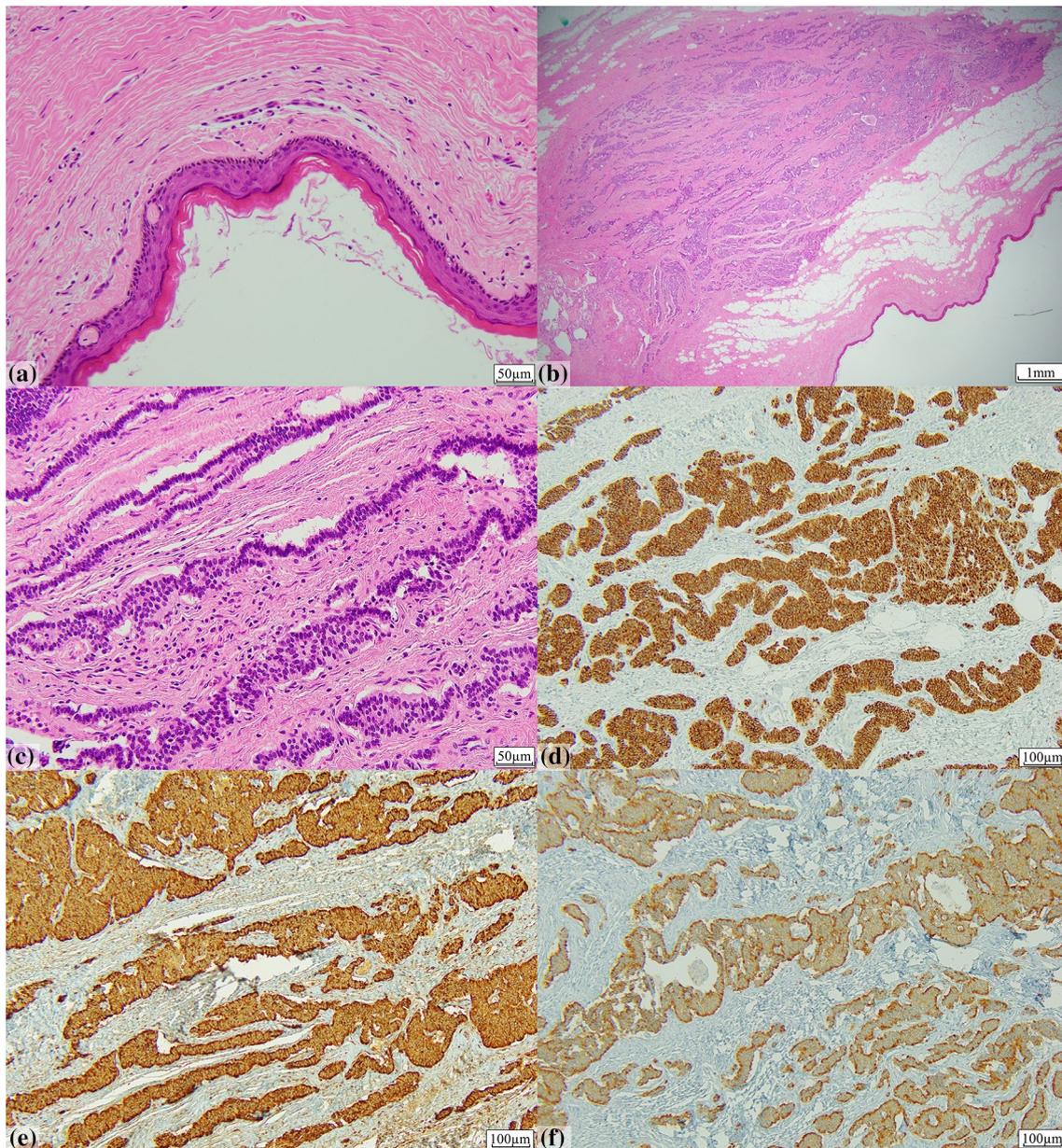


Fig. 2 Histology of the resected tailgut cyst. **a** Cyst lining with squamous and glandular areas. **b, c** Section of cyst with adjacent neuroendocrine tumour. **d** Tumour cells strongly positive for Cytokeratin

A1/3, e Tumour cells showing positive chromogranin and **f** synaptophysin stains showing neuroendocrine differentiation

in three and NET in one. Two of the four patients with malignant cysts had died in the follow-up period. Rates of malignant degeneration have also been reported at 6% [5], 9% [15], and 21% [6] in other studies. On the other hand, the largest series in the literature from Hjermsstad et al. [2] reported 51 patients with tailgut cysts and only 1 of them (2%) was accompanied with adenocarcinoma. Given that the range of malignant degeneration has been reported from 2 to 21%, studies have concluded that tailgut cysts should be removed because of the risk of malignant transformation, even if the patient is asymptomatic from the cyst [3].

However, the incidence of non-diagnosed asymptomatic cysts in the general population is unknown, and thus, it would be impossible to accurately estimate the risk of future malignancy from a tailgut cyst.

Pre-operative diagnosis of NET in a tailgut cyst can be challenging. Pre-operative biopsy was utilised in some, but not all studies, to confirm the presence of NET; these cystic lesions must be carefully sampled, as they can be quite large, and a malignant neoplasm may be some distance from the benign portion of the lesion [12]. Imaging is important for pre-operative diagnosis of tailgut cysts

to determine an optimal surgical approach and extent of resection [10, 16]. CT scan has been the most common first-line imaging modality in our review. CT can be used to demonstrate a well-circumscribed, presacral cyst containing water, or soft-tissue density and calcifications. In cases of malignant transformation, CT can reveal a loss of defined cyst margins as well as involvement of neighbouring structures [16]. In comparison, MRI scanning offers clearer visualisation of the multilocular appearance of the tailgut cyst together with improved detection of smaller peripheral cysts [10]. More importantly, it allows for better tissue characterisation because of its high-contrast resolution between different tissue compartments [17]. MRI also helps to determine solid tissue components (and thus potential neoplasm) within a tailgut lesion [18]. Sagittal MRI images offer an optimal view of the relation of the mass lesions to surrounding bony structures and the rectum. Thus, by offering multiplanar capabilities and good tissue contrast, MRI images are superior for the pre-operative planning for presacral cysts [15].

Conclusion

Incidental finding of malignancy in a tailgut cyst is rare. We present an uncommon case of NET in a long-standing tailgut cyst. The risk of malignant transformation or incidental finding occurring in unresected tailgut cysts is not negligible, and patients who are keen for a non-operative option should have a thorough discussion of their risk of missing a malignancy. The decision as to offer upfront resection or observation is dependent on the presence of any adverse features of malignancy on imaging, large size causing compression or obstruction of vital organs, and rapid progression in size. Pre-operative imaging with a pelvic MRI would offer better pre-operative detection of any malignancy in a tailgut cyst and also for surgical management planning. There is currently no specific recommendation as to the most appropriate interval follow-up time. We would suggest an initial 3 months repeat imaging, then yearly if there is no significant progression clinically and radiologically.

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Compliance with ethical standards

Conflict of interest Authors Alice Lee, Thomas Surya Suhardja, Thang Chien Nguyen and William Meng-Keat Teoh declare that they have no conflict of interest.

Human rights All procedures followed have been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

Informed consent Informed consent was obtained from all patients for being included in the study.

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