

Tailgut cyst: An uncommonly diagnosed common entity - A case report and review of literature

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ABSTRACT

Tailgut cysts (TGC) though relatively common are rarely diagnosed and documented. Most of the times, they are misdiagnosed and mismanaged. When malignancy develops in TGC it is rarely attributed to it as it is undiagnosed earlier and later on there may not be any evidence left behind. We report one such case which was diagnosed though not preoperatively. Review of literature reveals only few such cases after the initial description by Hjerstad and Helwig in 1988. Surprisingly most of the reports especially the larger case series are by radiologists. Surgeons and pathologists have contributed very few cases. This case report is aimed at increasing the awareness about this entity so that it will be suspected, investigated properly and treated before development of complications like recurrent fistulae, ischioanal abscesses which increase the morbidity and life threatening complications like malignant change.

Key words: Tailgut cyst, retrorectal cystic hamartoma, developmental cysts, retrorectal space

Introduction

Tailgut cysts, also known as retrorectal cystic hamartoma are rare congenital cysts that arise from the vestigial remnants of post anal hindgut. [1, 2] It is believed that the primitive hindgut extends into the tail of the embryo which is actually present at 8mm stage or 35 days gestational age. This extension being caudal to the future anus is known as post anal gut or Tailgut. It completely regresses by the gestational age of 56 days or 35mm stage. If it does not regress completely, the remnants may develop into Tailgut cyst [1, 2] which may be present congenitally [1] and may be identified or may become symptomatic even in neonatal period. [2, 3] However most of the patients are in their middle age. Almost

always TGC are located in the retrorectal or pre sacral space. Most of the patients are middle aged women who visit the outpatient department for chronic constipation.

Case Report

A 35 year old female reported to the outpatient department of Gynaecology of Bharati Hospital, Sangli, with chief complaints of retention of urine of 4 days duration. She also complained of dysmenorrhea for 2years and constipation off & on for last 3 years. She was para 4 with a history of first two FTNDs and last two LSCS. The last child birth was 10 years ago. During her last pregnancy she was investigated for difficulty in passing urine at 5 months gestation when a large encysted pelvic mass was identified at

USG examination. The mass was then aspirated & turbid fluid obtained. The fluid was found to be negative for AFB & the culture was also negative. Later on the cyst was biopsied and was reported as squamous epithelium lined cyst, having no evidence of tuberculosis or malignancy. At full term she underwent elective LSCS. After the childbirth she did not go for follow up to the surgeon probably because her symptoms were relieved. Then she gradually started having the pressure symptoms again with increasing severity for which she came to our hospital. As her previous symptoms were related to her pregnancy she attended the gynaecology OPD where she was investigated. The per abdominal, per vaginal, per speculum & per rectal examination findings were confirmed by CT scan with contrast which revealed a 15x20cm size cystic mass posterior to uterus, occupying the vaginal cavity posteriorly and compressing & displacing the rectum and urinary bladder anteriorly and to the right. After routine preoperative work up including CEA (Carcino Embryonic Antigen) which was normal, a laparotomy was planned with a tentative diagnosis of ovarian cystic mass. A panhysterectomy was performed and the surgeon was called as the cyst was found to be in the retrorectal region. The surgeon could do only partial excision of the cyst with marsupialization of cyst wall. A negative suction drain was kept. The post operative period was uneventful & the patient was discharged on 9th post operative day. At discharge the patient was counseled for total surgical removal of the cyst. Follow up examination one month after discharge revealed fluid collection within the cyst & the patient was again told to get it completely

excised. However she hasn't come for surgery until now, probably because she is temporarily relieved of her symptoms. Panhysterectomy specimen along with part of a thick cyst wall was received in the surgical pathology department. Except for a small endometrial polyp and hilus cell hyperplasia in bilateral ovaries the panhysterectomy was unremarkable. Microscopic examination of the cyst wall showed a non keratinizing stratified squamous epithelium lined thick fibromuscular cyst wall. The smooth muscle was arranged in disorganized manner having no resemblance to intestinal wall. There were no skin adnexal structures, no teratomatous elements, no myenteric nerve plexuses. There was no evidence of endometriosis or malignancy. (Fig.1) In view of these findings a histological diagnosis of Tailgut Cyst was offered.

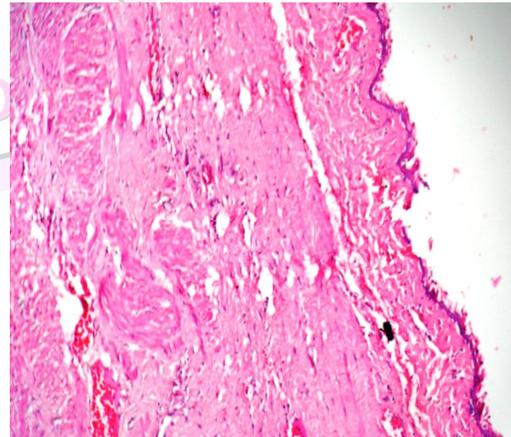


Fig. 1 Microphotograph showing cyst wall with stratified squamous lining and underlying muscle bundles

Discussion

Retrorectal space is a potential site for various developmental cysts like demoid, epidermal, duplication, Neurenteric and Tailgut cysts. Various other neoplastic and non neoplastic mass lesions can also occur in this region. These are cystic

sacrococcygeal teratoma, anterior sacral meningioma, anal duct or gland cyst, rectal leiomyosarcoma, extra peritoneal adenomucinosis (pseudo myxoma retroperitonei), cystic lymphangioma, pyogenic abscess, neurogenic cyst, sacral chordoma & sacral neurilemoma with cystic degeneration.

Tailgut cysts (TGC) are most often found in the retrorectal space. However case reports of Tailgut cyst outside this space are on record. Most common sites other than retrorectal space are around the perineum – sacrococcygeal subcutis, [4] perianal region, [1, 4] ischioanal fossa, [5] anorectal junction & anal sphincter. [6] Tailgut cyst of these locations usually have some connection with retrorectal space which may be in the form of a fibrous attachment with coccyx or lower sacrum. Extension of retrorectal TGC into the sacrococcygeal subcutis has been reported. [7] Far off locations like thigh [4] and perirenal space [1] are also on record. Rarely the Tailgut cyst may be situated prerectally. [1, 8]

About half of the patients may remain asymptomatic. Females are 3 times more commonly affected than men. This may be because females during pregnancies undergo repeated ultrasound examinations during which the TGC may be identified, or because of the TGC women may develop various gynecologic and obstetric problems for which they may be investigated like our patient. In women it can be mistaken for ovarian cyst. [2, 9] both by the radiologist/sonologist and by the gynecologist as in our case. Obstruction of birth canal & need for LSCS may be the presenting symptom. Though the usual presenting symptom is chronic constipation, other pressure symptoms like, low back ache,

painful defecation, urinary frequency, urinary retention, per rectal bleeding, sciatic pain, dystocia and dysperunia may be present. The TGC are usually of about 3.5cm size however cysts as big as 15cm have been reported. [5] In our case it was even bigger i.e. 20cm. If untreated complications like ischioanal abscess, fistula in ano, internal or per rectal bleeding, secondary infection and the most dreadful development of malignancy may occur. Adenocarcinoma is the commonest type of tumour, next common being carcinoid &/or neuro endocrine tumour. [6, 9, 10] Other malignant tumours developing in TGC are endometrioid carcinoma, adeno squamous carcinoma, squamous cell carcinoma & sarcoma. [1] Positivity for CEA & CA19.9, p⁵³ mutations & dysplasia-carcinoma sequence similar to colo rectal adenocarcinoma has been suggested. [1] Complete surgical removal by posterior approach or combined anterior and posterior approach is recommended. [2, 5] Diagnosis is made usually by radiological examination particularly by MRI. [2] TGC can be mistaken for adnexal mass by trans abdominal USG like in our case. Histopathological features that help differentiation of TGC from other developmental cysts are mentioned in Table 1.

A diagnostic biopsy is not advised as it may not be helpful in differential from other developmental cysts, malignancy may be missed and it may result in spillage and seeding of tumour cells. [1] MRI is a helpful technique to define extent of the mass its relationship to surrounding structures and also to demonstrate possible complication in order to choose the best surgical approach. Though MRI is the best

investigative modality for diagnosis of TGC, differentiation from other developmental cysts may be difficult especially if the TGC is unilocular. Absence of fat, location away from anus and

absence of communication with either rectum or thecal sac, rule out the diagnosis of dermoid cyst, anal gland cyst, rectal duplication cyst and anterior meningocele respectively on MRI study.

Table 1: Developmental cysts in Retrorectal space ^[11]
(Modified from Ref No 11)

	Epidermoid/ Dermoid cysts	Rectal Duplication Cysts	Retrorectal Hamartoma/ Tailgut cyst
Gross	Unilocular/ multi locular	Unilocular	may be Multilocular ± solid areas
Contents	Clear fluid/ dense muddy or fatty material	No contents/ mucoid secretions	Mucoid material
Cyst Lining	Squamous epithelium with/ without skin adnexa	Colonic/ Gastric/ Respiratory epithelium Presence of crypts, villi and muscularis mucosa	Squamous/ Transitional/ Glandular epithelium/ combination of these
Smooth muscle wall	No	Yes, Organized, Recapitulates muscularis propria	Yes, Disorganized bundles
Other Findings	---	Continuity or contiguity with rectum, Resemblance to rectum - two layers of muscle coat & mucosa & ectopic gastric or pancreatic tissue	Granulomatous or FB giant cell reaction

To conclude, we feel awareness of this entity, proper preoperative evaluation and total excision of the cyst help in prevention of complications which may cause lot of morbidity and mortality.

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