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Case Report

A case of adenocarcinoma in a tail gut cyst and review of literature

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Introduction

The tail gut cysts (TGC) are extremely rare benign congenital lesions arising from the persistent remnants of postanal gut and occur in retrorectal/presacral region.¹

In these as such rare benign lesions, malignant transformation is extremely rare, and we report such a case of tail gut cyst with adenocarcinoma arising from its mucosal lining, with its management, adjuvant therapy and follow up guidelines.

Case report

A 32-year-old male patient with an uneventful past medical and surgical history, presented with swelling over the intergluteal cleft associated with pain of 1-month duration.

He was diagnosed with gluteal abscess for which incision and drainage was done. Around 1 liter of clear mucoid material was drained. Following surgery mucopurulent discharge from the operated site continued. He reported to our hospital, MRI scan of pelvis showed a large well defined cystic lesion in presacral region measuring 9 cm × 12 cm × 19 cm which was suggestive of presacral complex cystic mass possibly a tail gut cyst (Fig. 1). Surgical excision of the cyst through a transsacral approach (Kraske's approach) was done (Fig. 2). Because of dense adhesions of the cyst wall with the surrounding tissue, the cyst could not be removed en bloc and had to be removed in a piece meal manner. A coccygectomy was added to enable better access post-operative period was uneventful. Patient did well and was discharged on seventh post-operative day.

Histopathological examination of the lesion showed a mucin secreting adenocarcinoma with villo-glandular pattern. Further immunohistochemistry evaluation of tissue block for markers showed it to be positive for CK7 in some tumour cells and diffusely and strongly positive for CK 20 and CDX-2 suggesting tumour to be of colorectal origin and excluded a metastasis. A post-operative F-18 FDG whole body PET-CT imaging showed an FDG avid lesion in the precoccygeal region with an uptake of SUV max 7.13 suggestive of post-operative tissue inflammation or a residual tumour. However CEA (4.46 ng/ml) and CA19-9 (1.2 U/ml) were within normal limits. In view of the piecemeal nature of excision, suspicious FDG uptake on the postoperative PET-CT scan and the patient being young, he was given 5040 cGy in 28 fractions over 47 treatment days with concurrent capecitabine followed by 8 cycles of

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Fig. 1 – Sagittal T2 weighted MRI scan through the pelvis showing a large high intensity multi-septate cystic tumour (black arrow) in the retrorectal space and its relation with the surrounding structures.

Fig. 3 – Sagittal CECT scan through the pelvis showing a small residual cyst in front of sacrococcygeal junction (black arrow) in the retrorectal space.

Fig. 2 – Rectum seen at the base after cyst excision.

FOLFOX 4. Repeat CT scan showed 5.5 cm × 4.5 cm × 3 cm retrorectal cyst (Fig. 3). CEA was 9.98 ng/ml. Excision of the remnant cyst was done through the same approach. Biopsy showed residual tumour in the excised cyst wall. Two-year post surgery, the patient is asymptomatic, having normal CEA with no recurrence on imaging.

Discussion

Often TGC is diagnosed and managed wrongly owing to its rarity and symptoms if present, mimicking inflammatory pathology.²

Complications can develop in TGC like secondary infection and malignant transformation. Amongst the malignancy, adenocarcinoma is the most common, followed by neuroendocrine tumour; other rare tumours include transitional cell carcinoma and sarcoma.³

A dysplasia-carcinoma sequence is often noted in cyst lining of TGC which is related to p53 gene mutation and therefore these tumours are often positive for CEA like other colorectal cancers.⁴

Recent studies have shown that the risk of malignant transformation in TGC is as high as 14% and not 2% as previously thought.^{1,3,5} Keeping this in mind, once the TGC is suspected, imaging should be done to especially look for the evidence of malignancy. Malignancy can be suspected on CECT or MRI if the wall of a cyst has an irregular, thickened look on the contrast image or T1 and T2 images. MRI is particularly useful in delineating soft-tissue planes and evaluating the presence or absence of bony invasion and nerve involvement which helps in surgical planning and hence MRI is the preferred imaging modality.⁶

The issue of whether to carry out biopsy preoperatively has remained controversial. Mathis et al. recommends a percutaneous preoperative parasacral biopsy, where the possibility of malignancy is high, however most surgeons claim that there is no role for biopsy of presacral lesions that are considered resectable. Moreover, there is significant concern about biopsy-related complications such as infection, haematoma and needle-track implantation.^{1-3,5}

Literature review of patients who have undergone excision of tail gut cyst harbouring adenocarcinoma has shown high recurrence rate within 5 months to 3 years.¹ Many a times, there has been previous attempts of drainage or excision as it is misdiagnosed as a benign inflammatory swelling or there may be an inadvertent cyst fluid leakage making tumour spillage unavoidable, moreover TNM staging is not possible in

Table 1 – Summary of cases of malignant transformation reported in literature.

References	No. of cases	Surgery	Adjuvant therapy	Follow up
Tampi et al. (2007)	17 ^a	+	–	Metastasis 10 months
Ballintyne (1932)	1		–	Died due to metastasis 8 months
Crowley (1960)	1		–	LR after 12 months
Hjermstads (1985)	1		–	Died after 8 months
Maruyama (1998)	1		CT	Alive after 3 years 2 months
Graadt Van (1999)	1		50.4 Gy RT	Alive for 4 years
Sauer (2000)	1	Redo surgery		Alive after 1 year FU
Puccio (2003)	1	+	CTRT	Disease free for 4 years
Rogers et al. (2007)	1	+	–	LR 2 years
Jarboui et al. (2008)	1	+	Macdonald CTRT	Alive after 4 years FU
Zappa et al. (2009)	1			
Mathis et al. (2010)	3	+	–	Died by last follow up
Fujisawa et al. (2010)	1		–	
Chhabra et al. (2013)	1	=	–	18 months disease free survival
Rammeh et al. (2013)	1	Redo surgery for locally advanced recurrent tumour	50 Gy RT	Died due to LR after 10 months
Present report	1	+	Undergoing CTRT	
Total	27			

^a Total no. of adenocarcinoma reported in literature till 2007 including one from that report.

Fig. 4 – Proposed algorithm for the management of TGC associated adenocarcinoma.

view of its ectopic site. For these reasons, in some cases, adjuvant radiation therapy with or without chemotherapy has been employed with good outcomes^{7–10} (Table 1). However, in the present case even after adjuvant therapy, CEA was high and CECT showed residual cyst, implying inadequate excision of cyst with incomplete response to CRT and hence necessitating re-excision.

There is no general consensus on treatment standard nor are there any treatment guidelines for TGC-associated adenocarcinoma because of the very low incidence rate (only 26 reported till date). However, limited evidence of rapid deterioration has favoured aggressive surgical removal of TGC soon after their diagnosis. As TGC associated adenocarcinoma are extremely rare, hence no randomized controlled trial will be possible, and in the absence of evidence, our proposed algorithm may serve as a guideline for this rare entity (Fig. 4).

Conclusion

Adenocarcinoma in a TGC is extremely rare. Surgeon needs to keep in mind the possibility of retrorectal cyst in case of suspected gluteal or perianal abscess with atypical characteristic, as incision and drainage could lead to dissemination, if the cyst harbours adenocarcinoma. We have also discussed the proposed management algorithm of adenocarcinoma in a tail gut cyst as none exists in literature.

Conflicts of interest

The authors have none to declare.

REFERENCES

1. Akbulut S. Unusual cause of defecation disturbance: a presacral tailgut cyst. *Eur Rev Med Pharmacol Sci.* 2013;17(12):1688–1699.
2. Singer MA, Cintron JR, Martz JE, Schoetz DJ, Abcarian H. Retrorectal cyst: a rare tumor frequently misdiagnosed. *J Am Coll Surg.* 2003;196(6):880–886.
3. Hjermsstad BM, Helwig EB. Tailgut cysts. Report of 53 cases. *Am J Clin Pathol.* 1988;89(2):139–147.
4. Schwarz RE, Lyda M, Lew M, Paz IB. A carcinoembryonic antigen-secreting adenocarcinoma arising within a retrorectal tailgut cyst: clinicopathological considerations. *Am J Gastroenterol.* 2000;95(5):1344–1347.
5. Mathis KL, Dozois EJ, Grewal MS, Metzger P, Larson DW, Devine RM. Malignant risk and surgical outcomes of presacral tailgut cysts. *Br J Surg.* 2010;97(4):575–579. <http://dx.doi.org/10.1002/bjs.6915>.
6. Mouloupoulos LA, Karvouni E, Kehagias D, Dimopoulos MA, Gouliamos A, Vlahos L. MR imaging of complex tail-gut cysts. *Clin Radiol.* 1999;54(2):118–122.
7. Tampi C, Lotwala V, Lakdawala M, Coelho K. Retrorectal cyst hamartoma (tailgut cyst) with malignant transformation. *Gynecol Oncol.* 2007;105(1):266–268.
8. Zappa L, Godwin TA, Sugarbaker PH. Tailgut cyst: an unusual cause of pseudomyxoma peritonei. *Tumori.* 2009;95(4):514–517.
9. Chhabra S, Wise S, Maloney-Patel N, Rezac C, Poplin E. Adenocarcinoma associated with tail gut cyst. *J Gastrointest Oncol.* 2013;4(1):97–100.
10. Rammeh S, Ben Abdelkrim S, Khalifa MH, Letaief R, Mokni M. Adenocarcinoma arising in a tailgut cyst: a case report. *Pathologica.* 2013;105(6):346–348.