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# Neuroendocrine carcinoma arising in a tailgut cyst

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ABSTRACT

*INTRODUCTION:* A tailgut cyst, also called retrorectal cystic hamartoma, is a rare congenital lesion that forms most commonly in the retrorectal space. It is presumed to arise from remnants of early embryogenesis.

*PRESENTATION OF CASE:* The following report describes a unique case of a retrorectal cystic hamartoma in a 53 year-old French Canadian man with a history of low back pain. The tumour underwent malignant transformation into a well-differentiated neuroendocrine carcinoma three years after the beginning of symptoms.

*DISCUSSION:* This condition can be found at any age, but occurs especially among middle-aged women. Not only is it frequently misdiagnosed, but also several complications associated to the cyst have been reported such as infection and malignant transformation. This is why complete surgical excision of the tailgut cyst is currently recommended.



## 1. Introduction

Tailgut cysts, or retrorectal cystic hamartomas, are uncommon congenital lesions occurring most of the time in the retrorectal space and among middle-aged women [1,2]. They have also rarely been found in the perirenal [3–5] and the prerectal spaces [1,5,6].

This lesion is thought to origin from an incomplete regression of a true tail located caudally to the future anus during early embryogenesis [9]. Although the cysts appear with non-specific signs and symptoms, an early diagnosis is the best way to avoid malignant transformation of the lesion, which is a rare but a possible complication [7]. Indeed, the following case report describes a 53 year-old French Canadian man with malignant neuroendocrine transformation of a tailgut cyst in accordance with the SCARE criteria [8].

The following work

## 2. Presentation of case

A 53-year-old man with a history of kidney surgery for a congenital malformation and a family history of prostate cancer, complained of low back pain for almost three years. Initially, the pain was located in the lumbar area only in the seated position. However, six months prior to consultation, the pain progressed and was also felt while walking. The physical examination did not reveal any apparent mass but palpation of the sacrococcygeal articulation provoked some pain to the patient. Initial investigation with magnetic resonance imaging (MRI) revealed a mass of 1.9 × 1.8 × 2.7 cm anterior to the coccyx. The mass was heterogeneous with a cystic part. On the T1-weighted images, a hyperintense cyst was visualized. The results then suggested either a possible chordoma or an exophytic giant cell tumour. The computed tomography (CT) scan, done two months later, confirmed the results of the MRI, also showing a cleavage plane between the rectum and the mass. Moreover, no calcification was seen in it. The patient underwent a coccygectomy with *en bloc* posterior surgical resection of the mass with negative resection margins. The rectum was left intact since the tumor was not invading it.

The gross examination showed a resected specimen measuring 7.6 × 3.9 × 2.5 cm. The resected coccyx was 3.7 cm of height, 1.0 cm of antero-posterior diameter and approximately 2.7 cm of width. A multiloculated cystic lesion with many different epithelial elements was observed: mucinous, ciliated columnar, transitional and malpighian (Fig. 1.). Within the wall of the cyst, a tumoral proliferation of polygonal, cuboidal and columnar cells was identified with irregular borders. The cells had an abundant acidophil cytoplasm with a round-shaped nucleus containing one to two nucleoli. The mitotic rate was 4–5 mitoses per 10 fields. The immunophenotype was positive for chromogranin (Fig. 2A), and synaptophysin (Fig. 2B), and keratin Cam 5.2 (Fig. 2C) in all cells, and a focal staining

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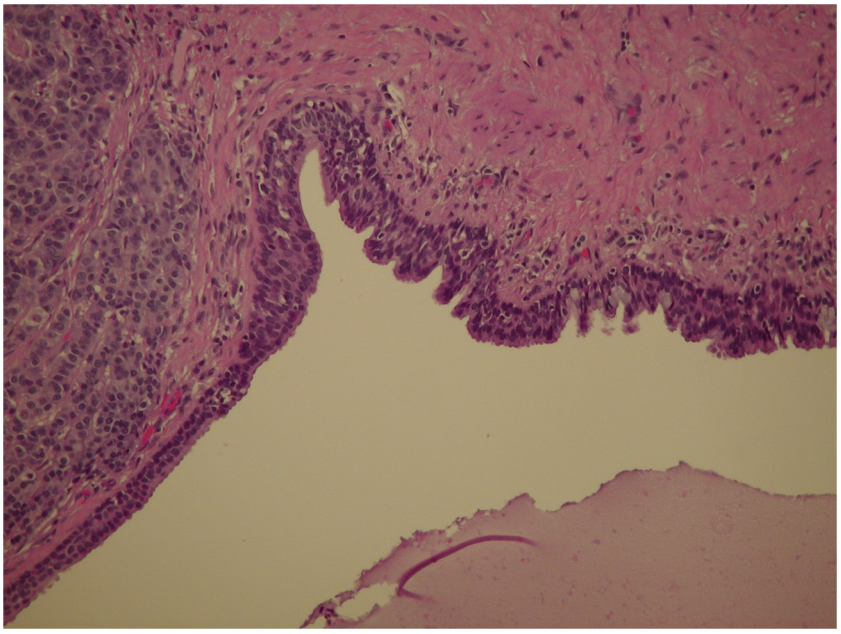

**Fig. 1.** The tailgut cyst is lined here by glandular epithelial cells including goblet cells, transitional cells and ciliated columnar cells. Tumor cells adopting a trabecular pattern of growth are seen on the left.

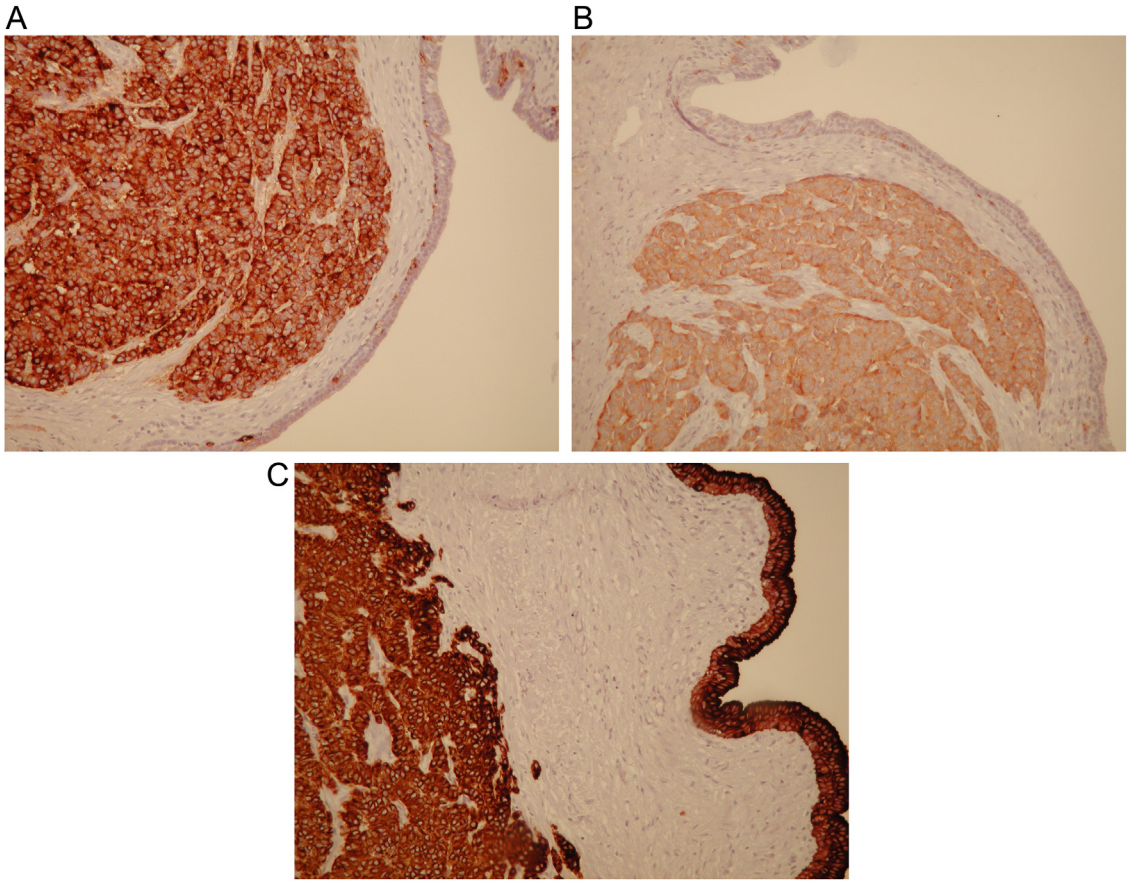


**Fig. 2.** A. Tumour cells expressing chromogranin. B. Immunohistochemically, the tumour cells were positive for synaptophysin. C. The tumour cells and epithelial lining of the cyst were strongly positive for keratin Cam 5.2.

was positive for keratin AE1-AE3 and CD56. The proliferation index (Ki-67) was 5–10%. The entire study of the specimen suggested a well-differentiated, grade 2/3, neuroendocrine tumour (NET) arising in a retrorectal cystic hamartoma.

The patient was doing well and had no recurrence of pain two months following surgery. No mass was palpated upon digital rectal examination and the seric chromogranin A (CgA) level was normal.