Case Report

Unusual Retrorectal Lesion

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Retrorectal lesions are rare entities. We report the complete clinicopathological details of an unusual retrorectal lesion composed of low-grade endometrioid adenocarcinoma and propose that this is a malignant change in pluripotent cells arising within a long-standing retrorectal tailgut lesion. To the best of our knowledge, this is the first case report of this malignancy in a retrorectal tailgut cyst. [Asian J Surg 2004;27(2):144-6]

Introduction

Retrorectal lesions are uncommon lesions that appear in the retrorectal (presacral) space, a potential space bounded anteriorly by the rectum and posteriorly by the sacrum. A variety of neoplastic and non-neoplastic conditions can occur in infants or adults in this region. The most common lesions encountered in children are teratomas, whereas in adults, chordomas and developmental cysts appear to be more frequent. We report the first case of an endometrioid carcinoma arising in a retrorectal tailgut cyst.

Case report

A 76-year-old woman was referred to the surgical outpatient clinic with perianal pain and tenesmus. On clinical examination, she was found to have a mass in the retrorectal region which was palpable on rectal examination. Computed tomography (CT) revealed the presence of two adjacent lobulated soft-tissue masses in the perirectal fat adjacent to the coccyx posteriorly and to a low-lying rectosigmoid junction anteriorly (Figure 1). Trucut biopsy via the perineal route, undertaken to assess risk and to plan the immediate treatment strategy, suggested a benign tailgut cyst. Subsequent to the biopsy, the patient had a myocardial infarction. At her 3-month follow-up, the patient elected for surgical removal of the lesion due to constant persistent pain. Due to the low level of the lesion, a Kraske’s approach was deemed most appropriate. The tumour was mobilized completely and did not appear to involve the rectum or the adjacent surrounding tissues. A good dissection plane was developed between the tumour and the sacrum, and the tumour was removed in its entirety with division of the S4 nerve root that appeared to be invaded by the tumour. The excision included the core biopsy needle track. The postoperative period was uneventful. No adjuvant therapy was given as the lesion was deemed to be completely excised. Laparoscopic examination of her pelvis revealed no evidence of endometriosis. No recurrence has been noted during 18 months of follow-up.

The gross specimen consisted of a mass of pinkish-tan firm tissue that weighed 54 g and, on cross-section, contained a fairly well-delineated solid pearly white gelatinous nodular mass measuring 6.5 × 3.5 × 4.5 cm. Histological analysis revealed the presence of an epithelial neoplasm that was divided into lobules by the presence of a fibrovascular stroma (Figure 2A). Complex labyrinthine papillary gland-like configurations with infoldings lined by flattened cuboidal, columnar and pseudostratified cells were observed. Superficial snouts and secretory cells reminiscent of endometrioid cells were seen associated with foci of squamous morels (Figure 2B). Areas of low cellularity with bland-looking epithelium contrasted with other regions showing increased cellularity with atypical epithelium and increased mitoses (Figure 2C).

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Immunohistochemical analysis showed that the cells in the lesion were positive for oestrogen receptor (ER) antibody and cytokeratin 7 (CK7). They were negative for carcinoembryonic antigen (CEA) and cytokeratin 20 (CK20). No evidence of endometriosis was found within the lesion.

Discussion

The human embryo undergoes many complex changes in the caudal region throughout early development. One of these events involves the appearance and disappearance of the embryonic tail associated with concomitant growth and regression of the hindgut. This tailgut segment lacks a muscular coat and consists of a tube that is usually lined with stratified cuboidal epithelium two to four cells thick, though variations such as intestinal or mucous epithelium are also observed. The tailgut atrophies and disappears completely in most adults. However, it is possible that vestigial remnants can persist and these are believed to be the origin of retrorectal hamartomas. Such retrorectal lesions are usually thin-walled, multicystic lesions lined with variable epithelium cell types, including stratified squamous, cuboidal, transitional, seromucinous and intestinal. These are more commonly reported in middle-aged women, in contrast to the retrorectal teratoma, which usually afflicts the paediatric population and is more commonly found in the anterior sacrococcygeal space. Other retrorectal cystic lesions found in this region include epidermal and presacral dermoid cysts, which are thought to arise from remnants of invaginated ectoderm, and duplication cysts of the rectum.

Malignant transformations, including adenocarcinomas and carcinoids, are known to arise in retrorectal tailgut cysts. These adenocarcinomas are usually moderately to poorly differentiated and are strongly positive for CEA. Complex papillae with nuclear crowding and atypia indicating a low-grade adenocarcinoma, as seen in our case, have not been described in these lesions. In addition, the immunohistochemical profile of this tumour (ER+, CK7+, CK20-, CEA-) supports a low-grade adenocarcinoma of the endometrioid type. The development of an endometrioid carcinoma in a retrorectal tailgut lesion has not been reported previously in the literature. The other most likely explanation of this carcinoma is that it originated from a focus of endometriosis. However, despite detailed histopathological search, no evidence to support this was found in the surgical specimen. There was no intraoperative evidence of endometriosis at the time of surgery and on postoperative laparoscopic examination of the pelvis. Detailed past clinical history did not reveal any supporting symptoms or history of endometriosis. Furthermore, the most common types of malignancy seen in conjunction with extraovarian endometriosis are clear-cell

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Figure 1. Computed tomography scan showing two adjacent lobulated soft-tissue masses (arrows) in the perirectal fat adjacent to the rectosigmoid junction.

Figure 2. A) Complex lobulated epithelial neoplasm (hematoxylin & eosin, low power). B) Superficial snouts and secretory cells reminiscent of endometrioid cells associated with foci of squamous morels (hematoxylin & eosin, medium power). C) Areas of increased cellularity with atypical epithelium (hematoxylin & eosin, medium power).
adenocarcinoma and adenosarcoma. Thus, we submit this as the first report of an endometrioid carcinoma arising within a long-standing retrorectal tailgut lesion.

**Conclusion**

Retrorectal lesions are rare and prove a challenge both in terms of diagnosis and surgical management. Malignant transformation within a retrorectal tailgut cyst is a rare but important complication to recognize. Such malignant foci can be low grade and focal in distribution. In this context, despite bland pathology at initial biopsy, we recommend complete surgical excision of these lesions for detailed histopathological evaluation.

**Acknowledgements**

The authors are grateful to Mr. Todd Reichert and Ms. Michelle Hesson in the preparation of Figures 1 and 2 for publication. This paper was originally presented as a poster at the 18th World Congress of Digestive Surgery, Hong Kong SAR, China, 8th to 11th December, 2003.

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