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Adenocarcinoma Arising in a Tailgut Cyst: Report of a Case

AKIRA MARUYAMA[1], KOJI MURABAYASHI[1], MASANOBU HAYASHI[1], HIDEAKI NAKANO[1], SHUJI ISAJI[1], SHINICHI UEHARA[1], TSUKASA KUSUDA[1], SHIGEKI MIYAHARA[1], AKINOBU KONDO[1], HIROSHI NAKANO[2], and TADASHI YABANA[2]

Departments of [1] Surgery and [2] Pathology, Yamada Red Cross Hospital, 810 Takabuku, Misono-mura, Watarai-gun, Mie 516, Japan

Abstract: We report herein the unusual case of a 66-year-old woman found to have adenocarcinoma arising in a tailgut cyst. The patient had been observed for 6 months following the discovery of a presacral cystic mass measuring 10 × 9 cm for which she had refused surgery. The serum tumor marker, carcinoembryonic antigen, became slightly elevated, and diagnostic imaging distinctly revealed a tumorous lesion with papillary projection into the cyst lumen. The cystic mass was then excised through the transsacral approach. The pathological findings were compatible with moderately differentiated adenocarcinoma arising in a tailgut cyst. This entity is extremely rare, and only six cases, including our own, have been reported in the English literature. Early complete excision is advised because it is almost impossible to determine for certain whether presacral cystic masses are benign or malignant prior to surgery.

Key Words: tailgut cyst, developmental cyst, presacral space, adenocarcinoma

Introduction

The tailgut cyst is an uncommon developmental anomaly in the presacral space that is frequently unrecognized, misdiagnosed, and mistreated. Adenocarcinoma arising in a tailgut cyst is extremely rare[1–6] and in this report, we present a case of this unusual entity, which to our knowledge is the first ever documented in Japan.

Reprint requests to: A. Maruyama, Daini-Iwasaki Hospital, 754 Isshinden-cho, Tsu-shi, Mie 514-01, Japan
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Case Report

A 66-year-old woman with no significant medical history was admitted to our hospital for the first time in January 1994 for investigation of bloody stools. Further evaluation revealed ischemic colitis of the transverse colon. In addition, a soft hemispherical extramural mass with a smooth surface was detected on the left posterior wall of the rectum by digital examination. A barium enema study demonstrated right-anterior displacement of the posterior wall of the rectum (Fig. 1). A computed tomography (CT) scan and T2-weighted magnetic resonance imaging (MRI) demonstrated a well-defined cystic mass with a smooth border measuring 10 × 9 cm in the presacral space (Fig. 2A). The patient refused to undergo surgery despite our recommendation. During 6 months of outpatient follow-up, she experienced gradual narrowing of the caliber of her stools with dull perianal pain, and in July 1994, she was readmitted to our hospital. Digital examination yielded almost the same findings as before. Laboratory studies showed values that were within the normal range, except for a carcinoembryonic antigen (CEA) level of 3.8 ng/ml, the normal being <2.5. While a CT scan disclosed no changes in the cystic mass at that time, T2-weighted MRI distinctly showed a solid component with a papillary projection into the cyst lumen (Fig. 2B). A pelvic arteriogram showed no tumor stain, and stretches of the distal branches were not encased. The mass was therefore diagnosed as a presacral cystic tumor, possibly having undergone malignant transformation, and dissection by the transsacral approach was performed. The coccyx and the fifth sacral vertebra were excised to gain access to the presacral mass. The thin-walled cystic mass, which compressed the rectum to the right and anteriorly, was adherent to the rectal wall, but was easily dissected away from it. During the dissection, no invasive lesions were noted in the surrounding structures. Because the cyst wall was slightly damaged at the

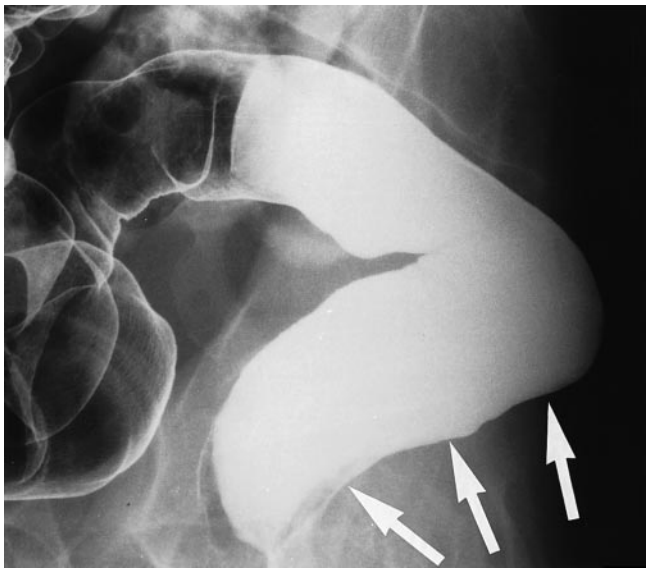

Fig. 1. Barium enema study showing right-anterior displacement of the posterior wall of the rectum (*arrows*)

site of retroperitoneal attachment, a total volume of 200 ml of fluid that had leaked from it was suctioned out. This reduced the size of the mass and facilitated complete removal. The specimen was a smooth-walled monolocular cyst measuring 9 × 7 cm which contained yellowish fluid and granular material.

Macroscopically, an elevated tumorous lesion, 3 × 2.5 cm in size, exhibiting papillary growth, was found attached to the cyst wall by a 3-mm-long stalk (Fig. 3). Frozen section examination revealed the lesion to be adenocarcinoma, but no evidence of malignancy was found in other parts of the cyst. Cytology of the contents of the cyst showed no evidence of malignancy. A drainage tube was inserted and the wound was closed.

Microscopically, the cyst was lined by a variety of types of epithelium, including stratified squamous, transitional, and columnar epithelium with goblet cells, but no evidence of malignancy was found (Fig. 4A–C). The lining did not have a complex structure with villi or crypts as seen in the normal bowel. Well-organized muscle fibers, myenteric plexus, serosa, and skin appendages were lacking in the cyst wall. The elevated lesion with papillary projection into the cyst lumen was confirmed to be a moderately differentiated adenocarcinoma (Fig. 4D). Its attachment to the wall, which lacked an epithelial covering, revealed granulation, but no malignancy. Consequently, a histopathologic diagnosis of adenocarcinoma arising in a tailgut cyst was established. Oral tegafur, 200 mg daily, was subsequently administered as adjuvant chemotherapy due to the elevation of the CEA level prior to surgery.

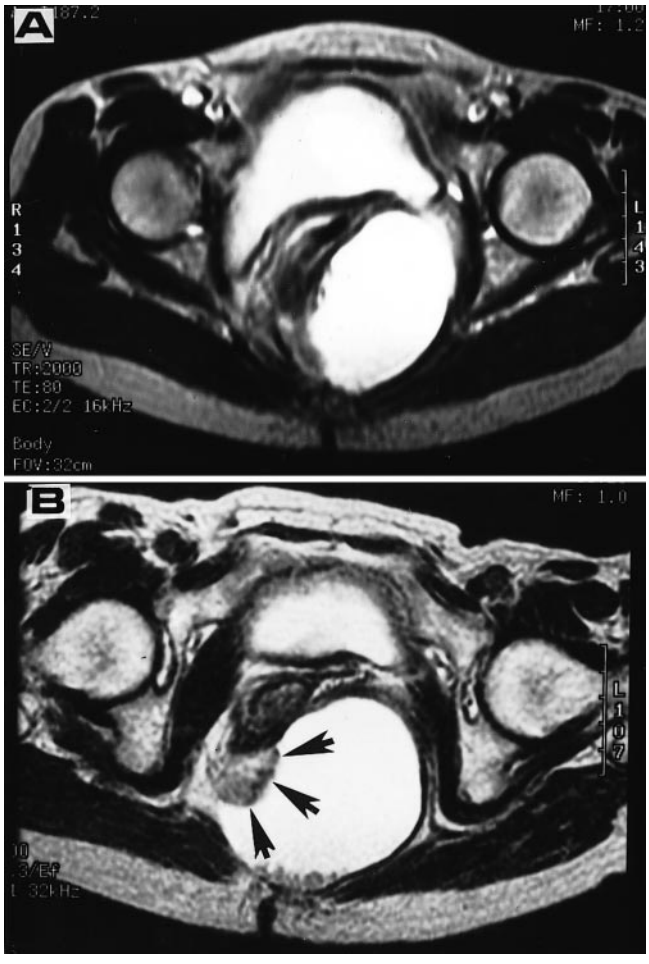

Fig. 2A,B. T2-weighted magnetic resonance imaging. **A** Appearance at the time of the first admission; **B** appearance 6 months later. A solid component with papillary projection into the cyst lumen was clearly visible (*arrows*)

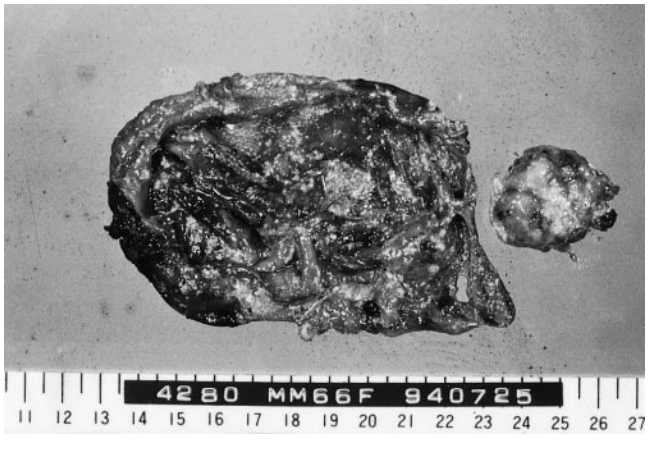

Fig. 3. Resected specimen. *Left*, A monolocular cyst containing yellowish fluid and granular material. *Right*, An elevated lesion, measuring 3 × 2.5 cm, in the cyst wall

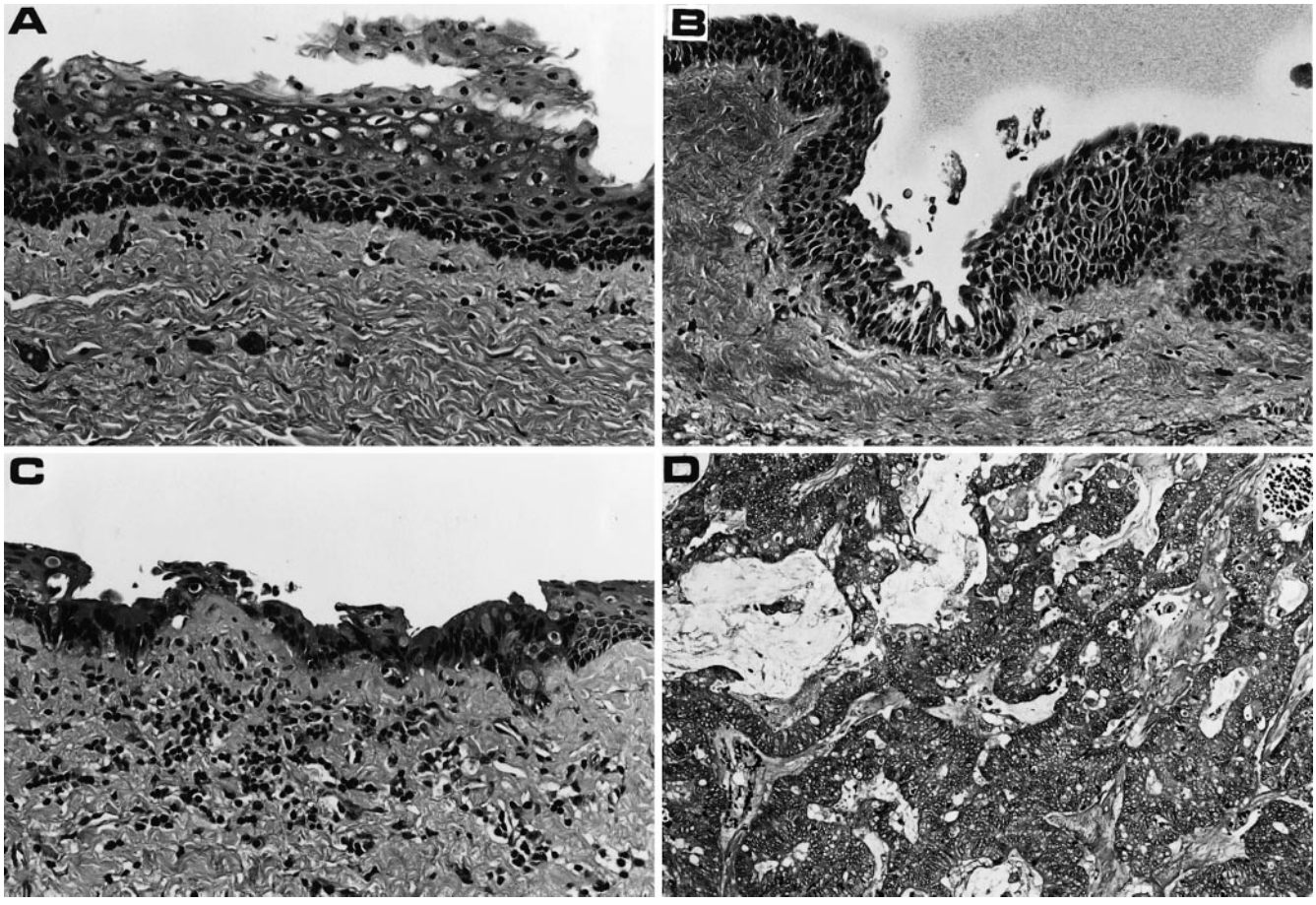

Fig. 4A–D. Microscopic findings. The cyst wall was composed of the following types of epithelium: stratified squamous (**A**), transitional (**B**), and columnar epithelium with goblet cells (**C**). The elevated lesion in the cyst wall was diagnosed as moderately differentiated adenocarcinoma (**D**). H&E: A, B, C, ×400; D, ×100

The patient had a satisfactory postoperative course, and has been well for the past 3 years and 2 months since her operation without any signs of recurrence.

Discussion

The presacral space, which consists of many types of embryonic tissues, is a potential site for the following variety of cysts and tumors: congenital, inflammatory, neurogenic, osseous, and miscellaneous other types, although the incidence of such cysts and tumors is low, especially in adults.[7,8] In 1953, Hawkins and Jackman[9] defined the term "developmental cyst" as a type of congenital cystic tumor in the presacral space due to some developmental error during the formation of the embryo. Such cysts include dermoid, epidermoid, and mucus-secreting cysts. They also suggested that mucus-secreting cysts originate from remnants of the neurenteric canal because they comprise both squamous epithelium and columnar mucus-secreting epithelium. On the other hand, in 1987, Hjermstad and Helwing[1] studied all retrorectal cysts diagnosed over a 35-year period at the American Armed Forces Institute of Pathology. They proposed the tailgut as the source of congenital cysts in the presacral space, and reported 53 cases of "tailgut cyst" with a review of an additional 28 cases documented in the literature. Their criteria specified that the cyst must be lined in part by columnar or transitional epithelium, but must not possess a myenteric plexus or serosa. This suggests that "tailgut cyst" is probably a synonym for "mucus-secreting cyst," but they doubted that the neurenteric canal was the source of the cysts because no neural elements were associated with them. Several authors have reported this entity since then.[6,8,10–14]

In Japan, only ten cases of mucus-secreting or tailgut cyst have been reported (8 in published papers,[15–22] and

the other 2 published as abstracts in congress reports). The female predominance of 7:3 and the average age of 39 years in Japan are almost identical to those stated in several other reports. All of the patients had symptoms, and about half had a perianal mass. Grossly, more than 50% of the lesions appeared multicystic, and the average diameter of all the lesions was 6.7 cm.

In view of the variety of pathologic diagnoses made in Hjermstad and Helwing's 53 cases and the fact that a correct histologic diagnosis was made in only 2 cases, it is expected that as the concept of tailgut cyst becomes more widely known, an increasing number of presacral cystic tumors may be found to be tailgut cysts.

Only five well-documented cases[2–6] of carcinoma arising in a tailgut cyst besides ours, including one case of carcinoma in situ (CIS),[4] have been reported in the English literature. Ballantyne[2] reported the first case of adenocarcinoma complicating a tailgut cyst in 1932. That patient developed local recurrences, lung metastases, and inguinal lymph node metastases, and died 8 months after removal of the cyst. The patient in the case reported by Crowley and Page[3] developed perianal recurrence, documented by biopsy, 12 months after removal of the cyst. The patient in Hjermstad's case report[5] died of her disease 8 months after abdominal perineal resection. Marco et al.[4] described a case of CIS developing in a tailgut cyst in which the patient was free of local or metastatic disease 20 months after surgery. Only a few cases of carcinoma in a tailgut cyst have been described in published reports, most of which had a bad outcome due to early local or metastatic disease, except for the single case of CIS. Conversely, the patient reported by Levert et al.[6] was only given adjuvant radiotherapy, and had a good outcome with 5-year survival, suggesting the effectiveness of adjuvant therapy. Although carcinoma supervening in a tailgut cyst is extremely rare, its possibility should be considered when dealing with developmental lesions of this nature. Despite recent advances in a variety of diagnostic methods such as ultrasound, CT, and MRI, which are able to be employed preoperatively, a precise diagnosis can only be made by pathological examination of the specimen after surgical removal. Therefore, early complete excision is recommended to avoid complications such as infection and recurrence that may occur even after removal of benign lesions,[8] and particularly to avoid malignant transformation.

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