

Case Report

Squamous cell carcinoma of the rectum, primary or metastatic?

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Introduction

Primary squamous cell carcinoma of the rectum is very rare. In this article we report a case of squamous cell carcinoma arising from an ovarian dermoid cyst presenting as a rectal mass with gastrointestinal symptoms. To the best of our knowledge, this clinical presentation has not been reported before. This article emphasizes the importance of applying the appropriate guidelines before assigning the diagnosis of primary colorectal squamous cell carcinoma, since the treatment and the management are different.

Case report

A 49-year-old female was transferred to University Hospital from an outside facility. She had a three month history of pelvic pain, hematochezia and difficulty defecating.

A CT scan revealed a necrotic colorectal mass most consistent with malignancy, which was seen eroding into a large teratoma within the pelvis. Rectal contrast was extravasating outside the bowel lumen and filling the teratoma consistent with a fistulous communication (Fig 1).

Her past medical history and family history were negative for malignancy. Because of her gastrointestinal

symptoms, she was referred to the gastrointestinal service for colonoscopy. On digital rectal examination a palpable mass was noted. The colonoscopy confirmed the presence of a partially obstructing ulcerated rectal mass (Fig 2) which was biopsied. The colonoscopy impression was consistent with primary rectal carcinoma. Microscopic examination of the biopsies revealed squamous cell carcinoma with adjacent normal colonic mucosa. The differential diagnosis included primary squamous cell carcinoma versus metastatic origin. Extension from the anal canal was also considered in the differential diagnosis. With the working diagnosis of teratoma with squamous cell carcinoma transformation extending to the rectum, the patient underwent total abdominal hysterectomy with bilateral salpingo-oophorectomy and low Hartmann resection of the rectum and sigmoid colon. The resection specimen revealed that the ulcerated rectal mass was directly adherent to the necrotic ovarian cyst (Fig 3). Microscopic examination supported the diagnosis of squamous cell carcinoma arising from ovarian dermoid cyst and extending to the wall of the rectum (Fig 4). The patient tolerated surgery well and was sent home in stable condition.

Discussion

Primary colorectal squamous cell carcinoma (SCC) is one of the rare malignancies of the gastrointestinal tract (1). It is part of the 2010 WHO classification, however pure squamous cell carcinoma is very rare and the incidence of (SSC) is far less than adenosquamous cell carcinoma (2). In 1919 Schmidtman described the first case of squamous cell carcinoma of the large intestine localized to the cecum (3). Since that time, less than 100 cases have been reported in the English literature.

No potential conflict of interest.

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Figure 1 Rectal contrast was extravasating outside the bowel lumen and filling the teratoma consistent with a fistulous communication



Figure 2 Colonoscopy view of the rectal mass eroding into the lumen

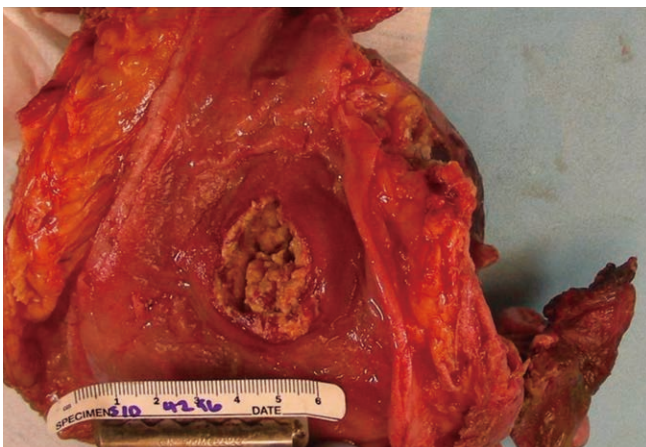


Figure 3 Colonoscopy view of the rectal mass eroding into the lumen

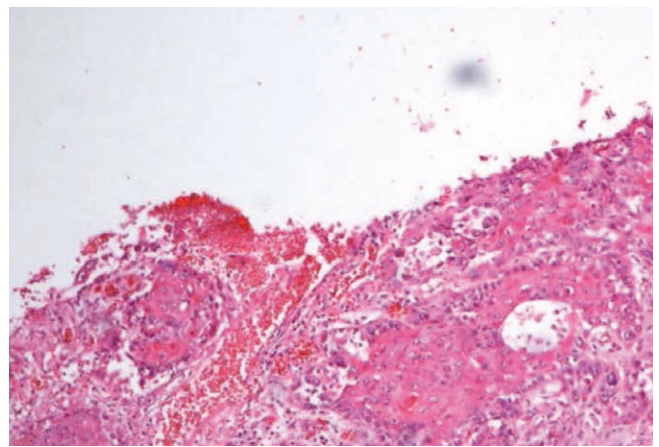


Figure 4 Invasive squamous cell carcinoma invading into the rectal wall

Of all cases of primary squamous cell carcinoma of the lower gastrointestinal tract, the rectum is the most frequent location for the disease, followed by the right colon (4).

Primary squamous cell carcinoma of the rectum affects individuals between the ages of 39 to 93, with a mean age of 57, years and is more frequent in women than in men (4). Squamous cell carcinoma can be seen in association with inflammatory and infectious processes involving the colon and rectum, such as ulcerative colitis, Schistosomiasis, Entamoeba histolytica and human papilloma virus (HPV) (4).

Since primary colorectal SCC are very rare. Williams et al. have suggested guidelines before making a definitive diagnosis of primary colorectal SCC, which include ruling

out the following entities: other primary sites, a squamous-lined fistula tract to the affected bowel and an extension of the tumor from the anal canal SCC. This can be established through careful clinical investigation and necessary radiographic images.

On the other hand mature cystic teratoma of the ovary is a common disease accounting for 10%–20% of all ovarian neoplasms (6). They are composed of well differentiated derivation of the three germ cell layers (endoderm, mesoderm, ectoderm). Complications of the mature cystic teratomas include torsion (16%), malignant transformation (2%), rupture (1%–2%), and infection (1%) (6,7).

Malignant transformation (MT) of an ovarian cystic

teratoma is rare and usually occurs in postmenopausal women (8). The most common type of malignant transformation is SCC arising from the squamous lining of the cyst, accounting for 80%–83% of cases, followed by adenocarcinoma (7%) and sarcoma (7%) (2). Primary SCC of the ovary is very rare (9). Ovarian squamous cell carcinoma might be associated with high risk human papilloma virus (10).

Most cases of ovarian SCC arise from a mature cystic teratoma, and are classified in the germ cell tumor category, although a few cases develop in association with endometriosis (9,11). The prognosis of MT is highly dependent on age, stage, and optimal cytoreduction, and there is no standard adjuvant treatment (12). Patients with SCC arising in mature cystic teratomas usually present with abdominal complaints (pain and mass) (6,10). These tumors grow slowly and cause minimal symptoms until they are very large or they become complicated (6).

The pathogenesis of the MT arising in ovarian MCT is not well understood. It is possible that since most MCTs are diagnosed during the reproductive age and MT is predominantly seen in the postmenopausal period, malignant transformation could be related to the long-term existence of non-removed MCT with prolonged exposure to various carcinogens in the pelvic cavity (8,10). Malignant transformation of MCT is usually diagnosed after removal of the tumor because it cannot be readily differentiated from an uncomplicated MCT or other ovarian tumors pre-operatively (8).

This case has a unique presentation, showing gastrointestinal symptoms, which was initially misleading. To the best of our knowledge squamous cell carcinoma arising from an ovarian dermoid cyst presenting as a rectal mass with gastrointestinal symptoms has not been previously reported. This case emphasizes the importance of the guidelines suggested by William et al, in the evaluation of patients with colorectal SCC. Since primary SCC of the colorectal are rare, other primary sites and an extension from the anal canal should always be considered.

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