On-line Case Report

A rare case of adenocarcinoma arising within a duplication cyst of the rectum: curative excision with 9-year follow-up

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This case report is novel in that it describes the successful excision of an adenocarcinoma arising within a rectal duplication cyst without evidence of disease recurrence at 9-years of follow-up.

Key words: Adenocarcinoma – Duplication cyst – Rectum – Curative excision

Intestinal duplications are congenital lesions and can occur in any region of the alimentary tract. Duplications may be tubular or cystic and occur most commonly in the midgut (50%), or foregut (36%) and least commonly in the hindgut (12%) with the rectum being the rarest site (5%).¹ In a review of the English medical literature, more than 70 cases of rectal duplication cysts have been reported.² Malignant change occurs rarely within a duplication cyst and was first described in the rectum of a 38-year-old woman in 1932.³ Nine additional cases have been described in which adenocarcinoma has arisen within a rectal duplication cyst.^{3–12} This case report is novel in that it describes the successful excision of an adenocarcinoma arising within a rectal duplication cyst without evidence of disease recurrence at 9-years of follow-up.

Case report

In November 1995, a 58-year-old female presented as an emergency with a discharging sinus from her right buttock, although she had been aware of a non-tender mass in the underlying tissue for one year. Her past medical history was unremarkable. Clinical examination

revealed a bloody gelatinous discharging sinus with an underlying 10-cm non-tender swelling. Digital rectal examination revealed a cystic mass located posterior to the rectal wall. No intraluminal abnormality was noted on rigid sigmoidoscopy. Due to the complex nature of the mass, a sinogram was performed. This demonstrated a large cavity within the right buttock that did not communicate with other structures. Subsequent magnetic resonance imaging (MRI) showed a posterior (presacral) mass measuring approximately 8 cm cranio-caudally and 10 x 6 cm transversely. There was no evidence of rectal wall involvement (Fig. 1).

Following discussion at a multidisciplinary team meeting involving the colorectal surgeon and radiologist, it was decided to attempt radical surgery. The patient was placed in a prone jack-knife position for the excision of the mass. Using a parasacral approach, wide *en-bloc* excision of the sinus track along with the cystic mass was undertaken. At surgery, there appeared to be no communication between the cystic mass and true rectal wall. In addition, there was no evidence of lymph node involvement in this mass. The entire tumour mass along with the lower segment of the sacrum below S4 and

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Figure 1 Coronal magnetic resonance imaging scan revealing a large presacral mass (S) with duplication cyst of the rectum (C).

coccyx were removed *en-bloc*. The patient made an uneventful recovery with full preservation of bladder and rectal function and was discharged home 6 days later. Histology confirmed a mucinous adenocarcinoma, infiltrating into the adjacent fibrous connective tissue but not into bone. The patient has been followed up at regular intervals and she remains disease-free at 9 years.

Discussion

Intestinal duplications are rare congenital anomalies of the alimentary tract. Duplication cysts of the rectum are the rarest, accounting for 5% of all intestinal duplications in a combined analysis of 455 patients.¹ Duplication cysts of the rectum are lined by alimentary tract mucosa, but not necessarily that of the rectum. Their wall usually shares a common layer of smooth muscle with the adjacent rectum with which they may communicate.¹³ There are several theories that explain their aetiology, but the most widely accepted explanation is that duplication occurs when a diverticulum forms during the development of the intestine in the 8–9-week-old embryo.¹⁴ Although the exact incidence of carcinoma arising within rectal duplication cysts is unknown, Weitzel and Breed¹¹ have reviewed the world literature revealing 25 cases of rectal duplication with carcinoma

arising in three of these and estimated an incidence of 12%. A further seven cases of malignant transformation has been reported in adults but it is extremely rare.^{3–12}

Diagnosis of this rare condition remains very difficult as most patients remain asymptomatic. Carcinoma arising from duplication cysts of the rectum may present with one or a combination of symptoms, including rectal pain, rectal bleeding, altered bowel function, perianal fistulation, or a mass.^{2–11,15–18} Computerised tomography or MRI are equally effective as imaging modalities. MRI is particularly helpful when radical surgery is being contemplated as it provides much better definition of the pelvic anatomy. Endorectal MRI has been shown to have better resolution than conventional MRI in demonstrating the common layer of smooth muscle with the adjacent duplication cyst.¹⁵ [¹⁸F]-Fluoro-2-deoxyglucose positron emission tomography (FDG-PET) has not been used in the diagnosis of this rare condition, however.

Although Monek *et al.*² have reported the case of a 39-year-old female whose rectal duplication presented as a buttock mass, it did not undergo malignant transformation. This differs from our case report in which an adenocarcinoma developed in the duplication cyst of the rectum that initially presented as discharging sinus overlying a buttock mass. There has been no evidence to support the use of chemotherapy or radiotherapy in the

treatment of adenocarcinoma arising in a duplication cyst of the rectum. Radical surgery offers the only hope for cure. The use of percutaneous biopsy to obtain tissue for the diagnosis should be discouraged because of the increased risk of needle track seeding of cancer cells. Contrary to previous reports^{3,5-7} that suggest a very high local recurrence rate despite radical surgery, our patient is disease-free at 9-years of follow-up.

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