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Case Report

Unusual presentation of mucinous adenocarcinoma in a post fistulectomy case

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Introduction

Carcinoma within a long-standing fistula-in-ano is rare and may be defined by specific neoplastic involvement of the fistulous track in the absence of rectal mucosal carcinoma.^{1,2} Such tumors are most frequently well-differentiated and mucinous in type, presenting late because of their insidious slow growing nature. These lesions are often mistaken for a benign condition. Due to their association with a symptomatic perianal fistula, malignant transformation is often initially overlooked.^{3,4} We present such a case along with a review of the available literature concerning its aetiopathogenesis.

Case report

A 66 years old male patient was operated in 1972 for fistula-in-ano. In 1995 he was reoperated for recurrence. Now he came to

surgical OPD with complaints of painless swelling in the right gluteal region of 4 years duration. There was no history of inflammatory bowel disease. On examination the swelling was involving right gluteal region reaching upto right perianal region. Overlying skin was normal in appearance and showed scar of previous surgery (Fig. 1 Inset A). The swelling was cystic. The differential diagnoses considered at this stage were perianal abscess, granulomatous lesion and adnexal tumor. CT scan revealed a large multiseptated loculated cystic SOL measuring $13 \times 9.3 \times 10$ cm lying within the periphery of right gluteus maximus muscle. The margins of the septae were smooth. The lesion was bulging anteromedially into the ischiorectal fossa, upper end bulging into levator sling and lower end bulging into gluteal fat. No fluid filled level and no solid component was seen. There was no significant enhancement in the lesion, in the septae and wall (Fig. 1). Possibility given was benign cystic neoplasm. FNAC was done which showed few endothelial cells and neutrophils entrapped in the background of mucin like substance. Based on these findings an opinion of benign mucinous cystic neoplasm was given (Fig. 2 Inset B). The possibilities were benign adnexal tumor or colitis cystica profunda. The lesion was excised. Intraoperatively 550 ml of mucinous fluid was collected (Fig. 1 Inset-B). Grossly, the mass was multiloculated with smooth cyst wall and few thickened areas. Histopathology of cyst wall revealed dysplastic columnar epithelium with abundant mucin. On complete processing of specimen mucinous adenocarcinoma was detected (Fig. 2). On colonoscopic biopsy rectal mucosa showed high grade dysplasia. In view of above the individual was taken up for Abdominoperineal resection (APR). The APR specimen showed moderately differentiated

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Fig. 1 – CT scan. Multiseptated loculated cystic SOL within the periphery of right gluteus maximus muscle (Inset A – right gluteal swelling; Inset B – mucinous fluid collected intraoperatively).

mucin secreting adenocarcinoma in the perianal region pT3 N0 M0; Stage II; R0. The rectal mucosa showed high grade dysplasia with no evidence of invasive adenocarcinoma. This was followed by adjuvant radiotherapy and chemotherapy.

Discussion

Primary mucinous adenocarcinoma of the anal region is rare and only accounts for 3% of all the anal carcinomas. Most of these tumors are ectodermal in origin. They can be classified into mucosal and extramucosal (perianal). Perianal adenocarcinoma seems to fall into two groups, based on their association with either fistulae or remnants of anal glands. At present, no laboratory methods can distinguish between these two. The risk factors associated with anal and perianal carcinoma are chronic fistula or anorectal fistula with chronic inflammatory diseases such as colitis or Crohn's disease. Mucinous adenocarcinoma of perianal region with preexisting fistula-in-ano is very rare with less than 200 cases reported in the literature, mainly single patient reports. However, incidence of mucinous adenocarcinoma of perianal region in preexisting Crohn's disease is 0.7%. The origin of this tumor is controversial: whether it arises from anal fistula, anal duct or

a duplicated duct is not known. It is frequently associated with fistula-in-ano, lack of involvement of rectal mucosa, and a well differentiated histologic appearance. Some are associated with Crohn's disease.⁵ The occurrence of a carcinoma in association with a fistula is probably due to chronic inflammation, although the rarity of the condition precludes any definite assumption in regard to the etiologic relationship of the fistula and carcinoma.

Rosner in 1931 established three essential criteria to confirm that malignant transformation has occurred within a fistula. He stated firstly that the fistula must be present for 10 years to exclude the possibility that the malignancy predated the fistula; secondly, that there should not be tumor within the mucosa of the rectum or anal canal unless there is definite evidence that this is metastatic tumor; and thirdly that the opening of the fistula within the anal canal or rectum should not contain malignant tissue.⁶ In the patient presented, the length of the preceding history was more than 10 years, however the rectal mucosa showed high grade dysplasia. One possible explanation for this may be carcinoma has developed in anorectal mucosa which may have been entrapped in the perianal soft tissue due to repeated surgery. At times, it may be difficult in some cases to ascertain that the tumor is a complication of a long-standing perianal fistula. The important

Fig. 2 – (H&E, ×400) Photomicrograph shows diffuse infiltration of signet ring cells. Giant cell (arrow) reaction seen. (Inset A, H&E, ×100 – shows mucin pools; Inset B – FNAC, Leishman & Giemsa, ×40 – shows few endothelial cells and neutrophils entrapped in the background of mucin like substance).

clinical features of perianal mucinous carcinoma usually include a long history of anal fistulae, and a painful buttock mass associated with discomfort on defecation and occasionally a clear gelatinous discharge. Extensive buttock ulceration is common. Unlike rectal ampullary adenocarcinoma, this tumor characteristically does not involve the rectal mucosa and is associated neither with bleeding nor obstruction.

Due to the rarity and slow growth rate of perianal mucinous carcinomas and the relative prevalence of anal fistulae, this tumor is usually not discovered for a prolonged period of time. The lack of early specific symptoms to differentiate it from fistulae with abscess formation makes diagnosis difficult. Such lesions present in an innocuous manner and can be mistaken for the more common clinical condition of a benign perianal abscess or fistula. A high index of clinical suspicion is required to make the diagnosis of perianal tumors while assessing patients presenting with perianal inflammatory conditions. MRI can provide better diagnostic information on patient with this suspicious condition.^{7,8} Wide excision of the tumor with abdominoperineal resection is the surgical treatment of choice and can provide good long-term results in patients with localized disease.⁹ Selected patients may require adjuvant chemotherapy.¹⁰ Our patient required radiotherapy and chemotherapy.

Conflicts of interest

All authors have none to declare.

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