Case report

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Mucinous Adenocarcinoma Arising from Chronic Perianal Fistula - A Case report

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Abstract

Mucinous adenocarcinoma arising from chronic perianal fistula in ano is a rare entity [1]. It is often mismanaged as benign disease due to its presentation. At present no guidelines have been established concerning diagnostic and treatment strategies due to the rarity of the tumour [2].A high index of clinical suspicion and biopsy of fistulous tracts and abscesses are key to early diagnosis and prompt treatment. We present a case of a 45 years old gentleman who initially presented with history of chronic perianal fistula for 3 years. Several biopsies from the fistulous tract, perianal skin opening and sigmoidoscopy biopsy were suggestive of benign nature of the disease. In view of high index of clinical suspicion and the nonhealing nature of the fistula tract, he underwent abdominal perineal resection with pediculated gracilis myocutaneous flap. The final histopathology report confirmed perianal mucinous adenocarcinoma.

Keywords: Perianal; Mucinous Adenocarcinoma; anorectal fistula.

Introduction

The prevalence of mucinous adenocarcinoma arising from chronic fistula in ano is rare and the diagnosis is often challenging. The relation between both entities was first described by Rosser et al. in 1934, who reported seven cases of fistula that had undergone malignant transformation [2]. It is postulated that the occurrence of carcinoma in association with fistula is probably due to chronic inflammation. However, the rarity of the condition deters any definite assumption in regard to establishing the etiologic relationship of fistula and carcinoma [3]. Most often, diagnosis is delayed due to the absence of alarming symptoms. Digital rectal examination does not help in establishing the diagnosis of mucinous adenocarcinoma in comparison to rectal carcinoma. Most of the time there will be only induration on the side where the fistula is situated. Biopsies of the external opening of the fistula tract very often are also not conclusive. It is mainly due to the fact the tissue taken is very superficial and only reveals an inflammatory reaction when scarring and fibrosis are present [4]. Nevertheless, excessive mucoid anal discharge or an abscess with mucoid content should arouse suspicion [5]. At present there is no guidelines or consensus in managing this condition.

The aim of this article is to report a rare occurrence of perianal mucinous adenocarcinoma which was treated with surgery even though the initial biopsies obtained did not suggest malignancy.

Case Report

We present a case of a 45 years old gentleman who presented with history of perianal discharge and pain for the last 3 years. Patient was previously fit with no underlying comorbidities. He had multiple episodes of perianal abscess drainage at a district hospital over the course of 3 years. On initial examination, noted patient had multiple external openings which were discharging mucoid material and pus. Initial colonoscopy done showed raised irregular mucosa over the distal rectum and biopsy showed features suggestive of solitary rectal ulcer syndrome. Patient underwent incision and drainage with seton insertion. A repeat biopsy under general anesthesia was also inconclusive and suggestive of benign in nature. He was subsequently referred to our center for further management of his complex disease. Magnetic resonance imaging (MRI) of pelvis showed multi-loculated complex perianal abscess with fistula communication. He underwent another examination under anesthesia and multiple samples taken from the fistula tract were sent for biopsy. During EUA we noted defect at rectum above the anorectal ring which was directly communicating with ischioanal space. The perianal region was filled with mucoid material. However, the histopathology report was still inconclusive and he was becoming more debilitated. All tumour markers and infective screening performed were negative. Furthermore, the biopsy did not show any findings of inflammatory bowel disease or carcinoma. Computed tomography of the thorax, abdomen and pelvis only showed the same finding description as the MRI. After a detailed discus-

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sion with the patient, we finally decided for surgical intervention to relieve the patient of his misery. He underwent abdominal perineal resection with end stoma. The large perineal defect was closed with pediculated gracilis myocutaneous flap by the plastic surgery team. His post operation recovery was uneventful and he was discharged after 14 days post-surgery. The final histopathology results from the abdominal perineal resection specimen showed well differentiated adenocarcinoma with >50 % of extracellular mucin with no lymph node involvement out of 53 lymph nodes harvested. All margins were clear. Patient recuperated well after the surgery and now currently undergoing adjuvant therapy.

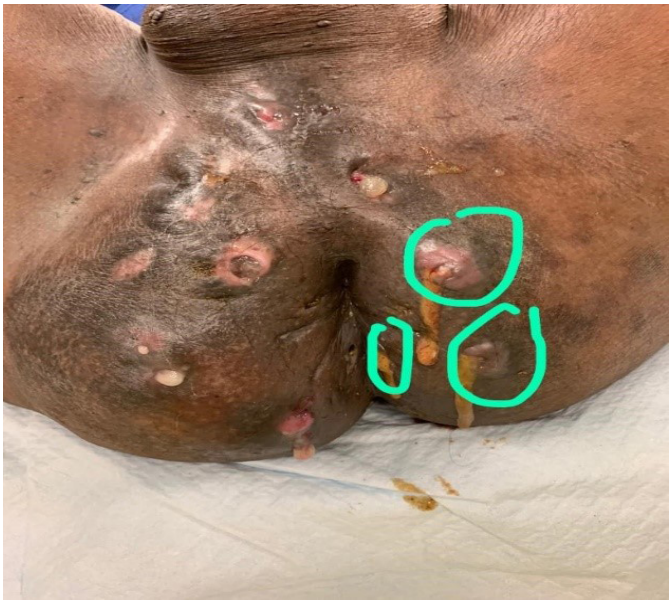

Figure 1: *Multiple external opening with pus and mucoid discharge.*

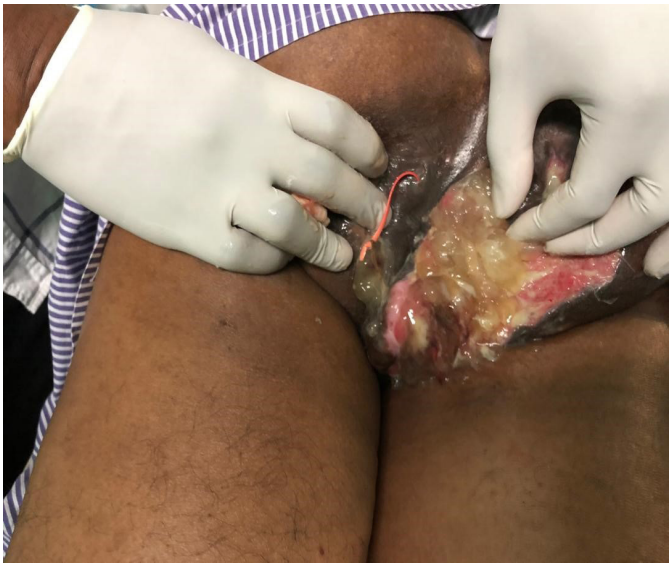

Figure 2: *Mucoid material over the previous incision and drainage site with seton in situ.*

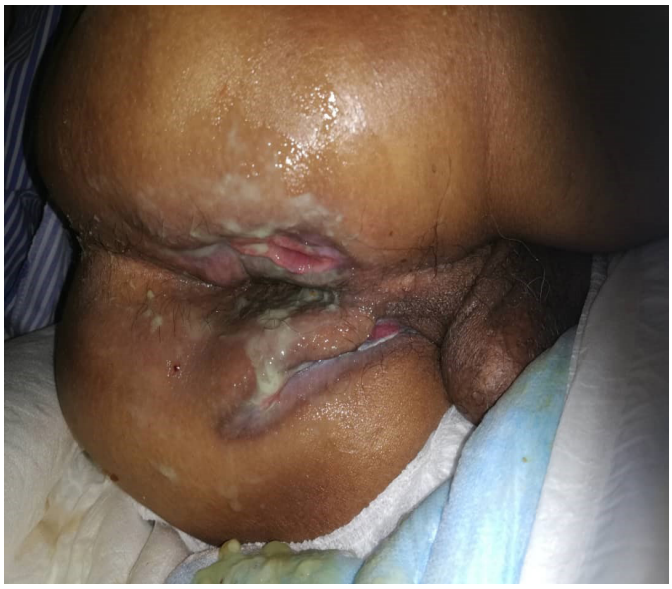

Figure 3: *Mucoid and pus discharge over the previous incision and drainage site.*

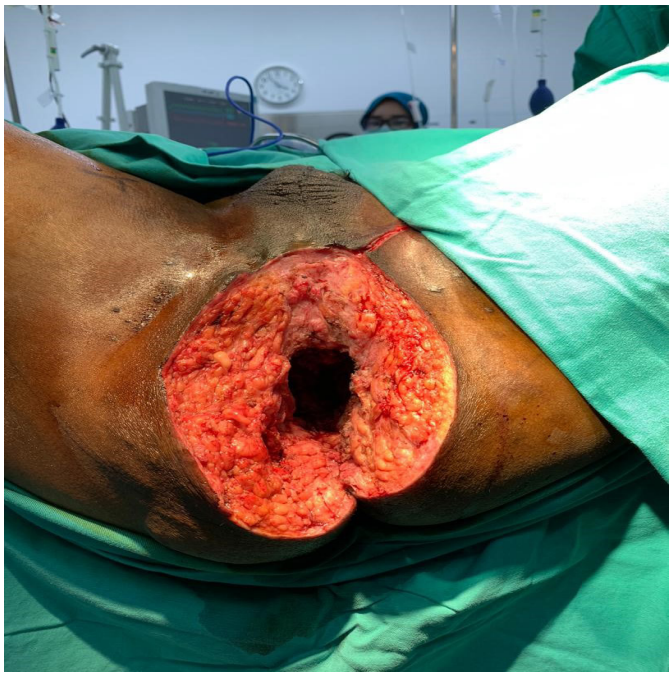

Figure 4: *Wide perineal excision done incorporating the previous incision and drainage site and area of induration.*

Discussion

Perianal mucinous adenocarcinoma arising from chronic perianal fistula are rare tumours and only accounts for 3-11% of perianal cancers [6]. It may arise de novo and present as a fistula, or it may arise from a long-standing perianal fistula or abscess cavity [6]. Most often it becomes a diagnosis dilemma in view of the absence of tumour within the lumen of bowel and slow growth of the lesion hidden within the ischioanal fossa or perineum [6,7]. Usually, it mimics benign presentation and most often clinicians fail to recognize it. The pathogenesis of the disease is still controversial [7]. It is usually associated with long standing fistula in ano and turns malignant secondary to

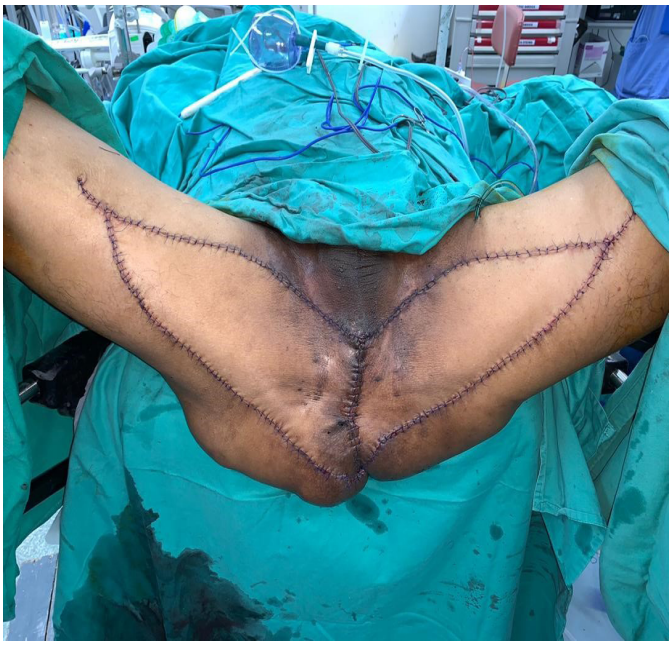

Figure 5: *Closure of the perineal defect with pediculated gracilis myocutaneous flap by plastic surgery team.*

chronic inflammatory changes [7]. Rosser postulated three possibilities describing the pathophysiology of this unusual entity; the fistula may precede the carcinoma by at least 10 years, the fistula opens inside the anal canal and outside the tumour, and any anorectal cancer should be an extension from the fistula harboring the carcinoma [7].

There are few key highlights points in our case which reflects the difficulty in diagnosing perianal mucinous adenocarcinoma. Firstly, the short duration of history of fistula which does not favor the arbitrary time period of ten years as suggested by Skir in 1948 [8]. The prior biopsies which were obtained were all inconclusive which resulted in depriving the patient of early neoadjuvant therapy. The decision to operate was based solely on clinical manifestations and not supported by negative biopsy reports.

We should have high index of suspicion in patients who have non healing chronic fistula in ano especially with mucinous discharge which fail to heal in spite of multiple attempts of surgery. Multiple biopsies of the suspected lesions are critical to early diagnosis and treatment due to the isolated areas of malignant transformation separated by inflammatory zones. Presence of extracellular mucinous lakes surrounded by well differentiated dilated tortuous glands, nerves and vessels in histopathology confirms the diagnosis [5]. However, in many cases definitive diagnosis is only verified by the final histopathological examination of the resected specimen [2].

MRI of pelvis plays an important role to establish the disease extension and to plan for the surgical treatment [2,4]. Abundance of mucin in the tumours gives rise to significant hyperintense signal on T2 weighted MRI images [2]. Despite that the interpretation of MRI occasionally can be challenging.

Once diagnosed, treatment should be radical. A proper multidisciplinary team approach should be undertaken prior to surgery to improve outcome. Patients usually will require neoadjuvant chemoradiotherapy for local control of disease before proceeding with definitive surgery. Hongo et al., in an 11-patient study, reported higher disease-free survival in patients who were given chemoradiotherapy prior to surgery [2]. Abdominal perineal resection with wide local excision is the preferred option as it guarantees a R0 resection and thus reduces the chances of local recurrence [9]. In selected cases pelvic floor reconstruction will be needed to cover the wide perineal defect after an ischioanal or extralevator APR [2,9].

In conclusion, perianal mucinous adenocarcinoma arising from an anorectal fistula is an uncommon malignant entity. Surgical decision should be made on clinical manifestation even if biopsy fail to provide conclusive findings. An aggressive and multimodal approach is key to achieving optimum patient outcome.

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