

# Perianal Mucinous Adenocarcinoma Diagnosed by Histological Study of Anorectal Abscess with Fistula

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## ABSTRACT

Perianal mucinous adenocarcinoma (PMA) is an oncologic rarity that poses a diagnostic and therapeutic dilemma for treating clinicians because there are few reported cases and an absence of definitive guidelines. We report a patient who had been treated with local surgery for recurrent perianal abscess with fistula for 3 years. Biopsy of the indurated tissue overlying his surgical scars revealed PMA. Neoadjuvant concurrent chemoradiotherapy followed by abdominoperineal resection was planned to address the locally advanced disease and ongoing sepsis. Our case is unique in that the fistula preceded carcinoma by only 3 years instead of the typical 10 years.

## INTRODUCTION

Perianal mucinous adenocarcinoma (PMA) is a rare tumor that can either present *de novo* or develop after a long-standing fistula-in-ano. PMA occurring in the context of a long-standing chronic fistula-in-ano is rare, with <150 cases, mostly single-patient cases, reported.<sup>1</sup> The absence of a tumor within the bowel lumen and the slow growth of a lesion hidden within the ischioanal fossa and perineum make early diagnosis difficult. Histological investigation of recurrent perianal abscess is important to aid early diagnosis.

## CASE REPORT

A 39-year-old Indian man presented with recurrent perianal abscess, for which incision and drainage procedures were performed 3 times in 3 years. During the most recent surgical treatment 5 months previously, a cutting seton was inserted. At consultation, he complained of anal pain, perianal induration, and purulent discharge from his perianal area. Examination under anesthesia revealed a fistula with an external opening at 4 o'clock and an internal opening at 6 o'clock, and the cutting seton was in place. The old surgical scars had a keloid-like appearance (Figure 1). Tissue biopsy of the indurated area overlying the scars revealed mucinous adenocarcinoma, with immunochemical histology positive for CK7, CK20, and CDX2. Colonoscopy did not reveal any pathology in the bowel lumen. Computed tomography of the thorax, abdomen, and pelvis showed a soft-tissue induration at the site of the fistula-in-ano, extending to the anal canal and left ischioanal fossa with possible involvement of the left levator ani muscle (T4) and multiple subcentimeter cervical, abdominal, and pelvic lymph nodes (N1) (Figure 2). No distant metastasis was found.

Neoadjuvant concurrent chemoradiotherapy (NACCRT), followed by laparoscopic abdominoperineal resection (APR), was planned. However, the surgery was done sooner due to ongoing perianal sepsis during NACCRT. The postoperative course was uneventful. Histological results confirmed extramucosal (perianal) mucinous

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**Figure 1.** A keloid-like appearance of the old surgical scar with the cutting seton visible.

adenocarcinoma with rectal circumferential margin involvement. Metastases to mesenteric lymph nodes were not detected (0/13). He then underwent further adjuvant concurrent chemoradiation (ACCRT) for 6 months.

## DISCUSSION

PMA is a very uncommon entity, accounting for only 3–11% of perianal cancers.<sup>2</sup> PMA may arise *de novo* and present as a fistula, or it may arise from a long-standing perianal fistula or abscess cavity. Few cases have reported anal ducts and glands as the sites of origin of perianal pathology. Some hypothesize that these ducts and glands may pierce the internal sphincter muscle before they extend to the perianal area in the ischioanal fossa, thus forming anal fistulas and perianal abscesses. Then, due to repeated friction, scarring, and



**Figure 2.** Computed tomography showing a soft-tissue induration at the site of fistula-in-ano, extending to the anal canal and left ischioanal fossa with possible involvement of the left levator ani muscle.

inflammatory reactions, the glands or ducts may transform into mucinous carcinomas, although there is no definitive evidence to support this theory. Another theory involves the deposition of malignant cells in the granulation tissue of a fistula from a proximal gastrointestinal cancer.

The clinical presentation of PMA can include perianal pain or itchiness, bleeding per rectum, bowel obstruction, or an ulcerative growth or palpable mass at the perianal region with bloody or mucoid discharge. Our case is unique in that the patient presented with a history of preceding perianal abscess and fistula of only 3 years' duration; previous case reports and case series had established a minimum preceding fistula duration of 10 years.<sup>3</sup> It is unclear whether the fistula was the source of the tumor or whether it was merely the initial presentation of the anorectal carcinoma. However, with the exclusion of other carcinomas with colonoscopy and computed tomography, we believe that the carcinoma in this case arose from a benign fistula.

The early diagnosis of PMA is difficult as the tumor has an indolent growth and remains hidden within the perianal region. Malignancy should be suspected in a chronic perianal fistula that shows early induration; has continued discharge, especially mucinous discharge; and does not heal after surgery. There may also be isolated areas of malignant transformation separated by inflammatory zones. Therefore, multiple biopsies of the suspected lesions are critical to early diagnosis and early treatment, particularly in patients who presented with recurrent perianal abscess or long-standing fistula-in-ano. Once diagnosed, treatment should be radical. Anal duct pathology, especially the known aggressive variety, has been managed with APR followed by ACCRT or NACCRT and then APR in view of its highly advanced nature with surrounding tissue invasion.<sup>4</sup> Although the role of NACCRT in the treatment of PMA has not yet been established, a recent case study has demonstrated good results in tumor regression and downstaging.<sup>1</sup>

PMA is an uncommon complication of chronic perianal fistula. Our report suggests that progression from a benign fistula-in-ano to PMA may be faster than previously described. A high degree of clinical suspicion and a histopathological confirmation are required to identify and diagnose this entity, particularly in an ulcerated or indurated area of the perianal region with a history of long-standing perianal abscess or fistula-in-ano.

## DISCLOSURES

Author contributions: All authors contributed equally to the manuscript. SL Jee is the article guarantor.

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Informed consent was obtained for this case report.

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