

Case Report

Appendiceal signet ring cell carcinoma masquerading as acute appendicitis: a case report

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ABSTRACT

Appendiceal signet ring cell carcinoma (ASRCC) is a rare and aggressive malignancy of the appendix. It often presents with nonspecific symptoms resembling acute appendicitis, making early diagnosis challenging. Prompt recognition and appropriate management are essential to improve outcomes. A 28-year-old gentleman presented with pain in the right lower abdomen for 2 weeks, associated with low-grade fever. Abdominal examination revealed tenderness and guarding in the right lower quadrant. Computed tomography (CT) of the abdomen revealed a retrocaecal appendix with severe acute appendicitis, appendicular tip perforation, and a peri-appendicular collection. The patient underwent laparoscopic drainage of the abscess with appendectomy. Histopathological examination of the specimen revealed signet ring cell carcinoma of the appendix. Further staging evaluation was performed, following which the patient underwent cytoreductive surgery comprising right hemicolectomy and regional lymphadenectomy, along with hyperthermic intraperitoneal chemotherapy (HIPEC). Histopathological examination established the diagnosis of appendiceal signet ring cell carcinoma following an initial presentation as perforated acute appendicitis. The patient subsequently underwent definitive oncological management with cytoreductive surgery and HIPEC. A comprehensive review of the available literature was also undertaken, focusing on the clinical presentation, histopathological characteristics, proposed pathogenesis, treatment strategies, and prognosis of ASRCC. Appendiceal signet ring cell carcinoma is an uncommon but highly aggressive malignancy that may mimic acute appendicitis, particularly in cases presenting with perforation or peri-appendicular abscess. Histopathological examination of appendectomy specimens is crucial for detecting unexpected malignancy. Early diagnosis, appropriate staging, and timely multidisciplinary oncological management, including cytoreductive surgery and HIPEC in selected patients, may improve clinical outcomes.

Keywords: Acute appendicitis, Appendiceal adenocarcinoma, Appendiceal signet ring cell carcinoma, Cytoreductive surgery, Hyperthermic intraperitoneal chemotherapy, Peritoneal carcinomatosis

INTRODUCTION

Acute appendicitis is the most common indication for emergency abdominal surgery worldwide and remains the leading cause of abdominal surgical intervention in the paediatric population.¹ Histopathological examination of appendectomy specimens reveals appendiceal malignancies in approximately 0.5%–1% of cases.² Among these, epithelial neoplasms account for nearly 0.2%–0.3%, while neuroendocrine tumors are identified in about 0.3%–0.9% of specimens.²

Primary adenocarcinoma of the appendix is a rare gastrointestinal malignancy, representing less than 0.5% of all gastrointestinal cancers.³ Within appendiceal malignancies, appendiceal signet ring cell carcinoma (ASRCC) is an exceptionally uncommon histological subtype, constituting approximately 4% of primary appendiceal carcinomas.⁴ Histologically, ASRCC is characterized by malignant epithelial cells containing abundant intracytoplasmic mucin, which displaces the nucleus to the periphery, producing the characteristic signet ring morphology.⁵

Signet ring cell carcinoma can arise in multiple organs; however, the stomach is the most frequent primary site. Consequently, primary ASRCC of the appendix is exceedingly rare and is associated with aggressive biological behaviour and poor prognosis.⁶ Owing to its infiltrative growth pattern and early metastatic potential, ASRCC is often diagnosed at an advanced stage.³

Clinically, ASRCC commonly presents with nonspecific symptoms that closely resemble acute appendicitis, most frequently right lower quadrant abdominal pain.² This clinical overlap makes preoperative diagnosis challenging, and the condition is usually identified incidentally following appendectomy on histopathological examination.⁷ At the time of diagnosis, many patients demonstrate regional lymph node involvement, peritoneal dissemination, or metastasis to adjacent organs, reflecting the aggressive nature of the disease.³

This case report and review of the literature aim to increase awareness of appendiceal ASRCC as a rare cause of acute appendicitis and to summarize current knowledge regarding its clinical presentation, histopathological features, pathogenesis, treatment options, and prognosis.

CASE REPORT

Patient information

A 28-year-old gentleman presented to emergency department with complains of pain in right lower abdomen for 2 weeks, intermittent, non-radiating, not relieved with medications. Also complains of fever for 1 week

associated with chills, low grade, not relieved with medications. The patient reported no altered bowel habits, weight loss, or any previous episodes of similar abdominal pain. He had no significant past medical history and no prior gastrointestinal surgeries.

Clinical examination

On physical examination, patient was well built and nourished. His vital signs were as follows: temperature 100.2 °F, blood pressure 130/80 mmHg, heart rate 89 beats per minute, and respiratory rate 18 breaths per minute. An abdominal examination revealed tenderness and guarding in the right lower quadrant, with a positive Rovsing's sign. Bowel sounds were present.

Ultrasound abdomen and pelvis showed mass in right lower abdomen in right iliac fossa with internal debris suggestive of Acute appendicitis with appendicular mass. Computed tomography (CT) of abdomen and pelvis showed retrocaecal appendix with severe acute appendicitis, appendicular tip perforation and peri-appendicular collection which is abutting the psoas muscle sheath with severe oedema of muscle (Figure 1A, B).

Management and follow-up

Patient underwent laparoscopic drainage of abscess with appendectomy. Intraoperatively, thick-walled gangrenous appendix with perforation at its tip with abscess collection of approximately 40-50cc pus noted retroperitoneally (Figure 2). Excised appendix specimen was sent for histopathological analysis.

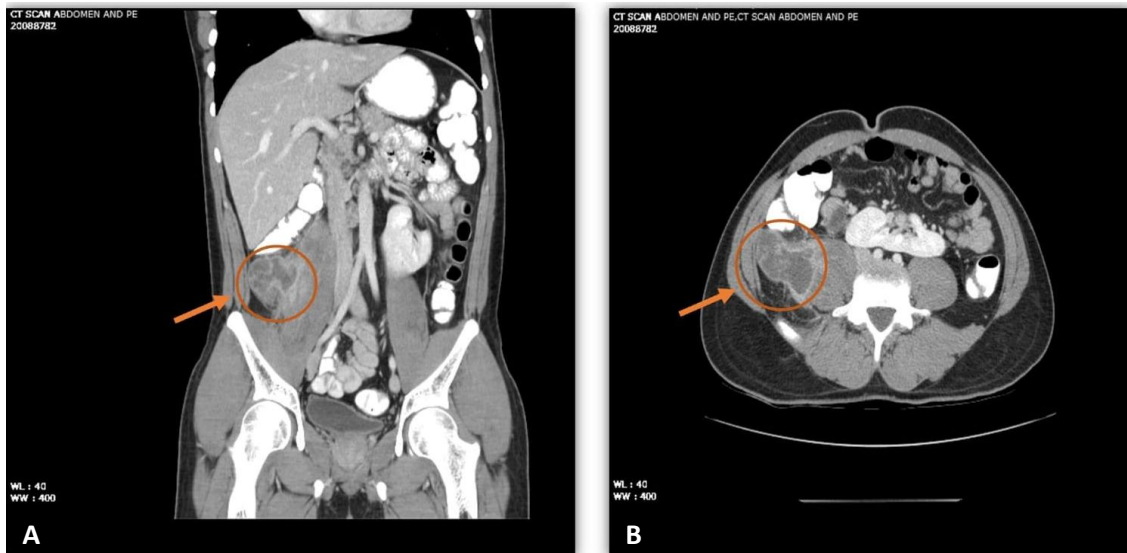


Figure 1: CT abdomen and pelvis showing severe appendicitis with peri-appendicular collection. (A) coronal view, (B) transverse view.

Histopathological analysis was reported as Signet ring cell carcinoma of Grade 3 (Figure 3). The tumor was classified as T3 N0 M0, indicating locally advanced but non-

metastatic disease, according to The American Joint Committee on Cancer staging system for appendiceal carcinoma.



Figure 2: Gross specimen of appendix with perforation at its tip.

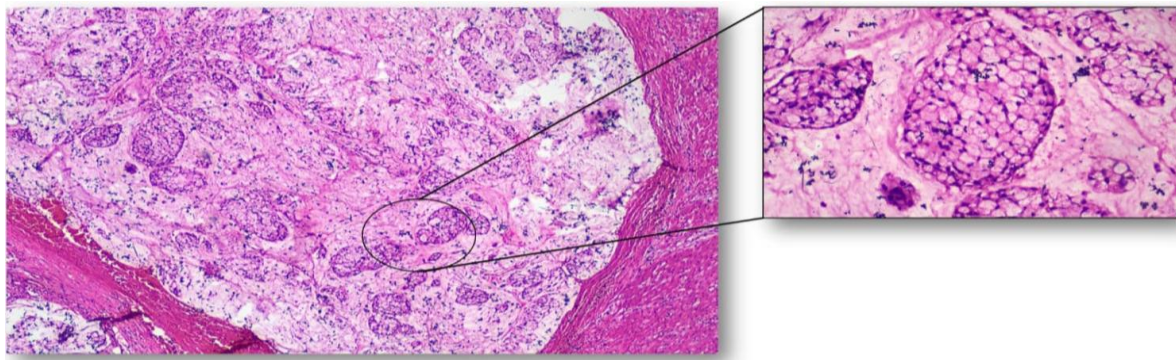


Figure 3. Histopathological examination with hematoxylin and eosin (H&E) staining of the appendix showing signet ring cells and extracellular mucin suggesting of signet ring cell carcinoma.

After diagnosis of Appendiceal signet ring cell carcinoma, patient underwent staging evaluation with CT chest, abdomen and pelvis. CT showed post operative changes at caecal base along with an enhancing lesion that appears adherent to the right psoas. Patient's tumor markers, including carcinoembryonic antigen (CEA) and cancer antigen 19-9 (CA 19-9) were within normal limits.

Patient underwent cytoreductive surgery (right hemicolectomy and regional lymphadenectomy) with Hyperthermic intraperitoneal chemotherapy and the histopathological analysis of the resected specimen showed residual signet ring cell carcinoma at base of caecum, with involvement of peritoneum and with pericolic lymph node metastasis. Patient's post operative course was uneventful and discharged on 6th day. Patient was being planned for adjuvant therapy with intravenous 5-fluorouracil (5-FU) due to the aggressive nature of signet ring cell carcinoma on follow-up.

DISCUSSION

Appendiceal adenocarcinoma arises from the glandular epithelium of the vermiform appendix and, despite its rarity, exhibits significant malignant potential. In most cases, diagnosis is incidental following appendectomy performed for suspected acute appendicitis, as observed in the present case.

The mean age at diagnosis for appendiceal mucinous adenocarcinoma is approximately 60 years, whereas ASRCC tends to present slightly later, with a reported mean age of around 62 years. Notably, ASRCC shows a marked female predominance, with a reported male-to-female ratio of approximately 1:11.³

The pathogenesis of signet ring cell carcinoma is believed to involve genetic alterations in pluripotent intestinal crypt epithelial stem cells, resulting in abnormal intracellular mucin accumulation and the formation of neuroendocrine

secretory granules.⁸ Accurate preoperative identification of appendiceal neoplasms is crucial for appropriate surgical planning in patients presenting with features of acute appendicitis.⁷ However, the lack of pathognomonic imaging findings often makes differentiation from uncomplicated appendicitis difficult.

Cross-sectional imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) may aid in suggesting an underlying malignancy. Imaging features suspicious for neoplasia include a heterogeneous appendiceal mass with wall enhancement, peri appendiceal fat stranding, and associated regional lymphadenopathy.⁹ In contrast, appendicoliths, intramural air, localized fluid collections, and focal thickening of the caecal apex are more characteristic of acute appendicitis¹⁰. Advanced age at presentation should also raise suspicion of an underlying appendiceal neoplasm.¹¹

Diagnosis of ASRCC is particularly challenging, as metastatic disease is frequently present at the time of detection when compared with other primary appendiceal tumors. Consequently, ASRCC is associated with a poor prognosis, especially in the presence of distant metastasis. McGory et al. reported distant metastases in approximately 60% of patients and a five-year survival rate of only 18%.¹² Even when localized, ASRCC demonstrates significantly lower survival rates than other histological subtypes of appendiceal malignancies.

The optimal management of ASRCC remains uncertain and should be individualized based on clinical and pathological factors. In early-stage disease confined to the mucosa, appendectomy alone may be curative.⁶ In advanced disease, right hemicolectomy with regional lymph node dissection is recommended.¹²

The role of adjuvant chemotherapy remains controversial; however, it may be considered in the presence of high-risk features such as perforation or invasion of adjacent structures. Current National Comprehensive Cancer Network (NCCN) guidelines recommend managing appendiceal adenocarcinomas similarly to colorectal cancer, including the use of 5-fluorouracil-based chemotherapy.¹³ Adjuvant regimens such as CAPEOX may offer a survival benefit in patients with signet ring cell carcinoma.¹⁴

For patients with metastatic disease, systemic chemotherapy is a reasonable option. Cytoreductive surgery combined with hyperthermic intraperitoneal chemotherapy (CRS/HIPEC) is considered the standard treatment for peritoneal dissemination from appendiceal malignancies.^{15,16} However, its benefit in ASRCC remains controversial due to the aggressive biological behaviour of this tumor subtype.

Although signet ring cell carcinoma is rare and typically shows a female predominance with a peak incidence in

patients over 60 years of age, our case is unusual in that it occurred in a 28-year-old young male.

CONCLUSION

In conclusion, appendiceal signet ring cell carcinoma is a rare and highly aggressive malignancy that often presents with vague and nonspecific clinical features, posing significant challenges to early diagnosis. Timely recognition and appropriate management are essential for improving patient outcomes, and optimal care requires a multidisciplinary approach. Imaging modalities, along with meticulous histopathological evaluation of the resected appendix, play a pivotal role in establishing an accurate diagnosis. Surgical resection remains the cornerstone of treatment for appendiceal malignancies, while adjuvant chemotherapy may be considered in selected high-risk cases. Clinicians should maintain a high index of suspicion for appendiceal signet ring cell carcinoma in patients presenting with features of acute appendicitis, particularly in those with atypical clinical presentations or unexpected histopathological findings.

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