



Case report

Primary adenocarcinoma of the appendix presenting with fresh bleeding per rectum: A case report

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ABSTRACT

Introduction: Primary adenocarcinoma of appendix is a rarely diagnosed malignancy accounting for less than 6% of appendiceal neoplastic lesions and less than 0.5% of all gastrointestinal malignancies. It is mostly diagnosed as an incidental finding after appendicectomy.

Case summary: An 81 year old male patient presented with bleeding per rectum in a background of previous rectal polyp, hypertension, diabetes and hypothyroidism. CECT of whole abdomen findings revealed thickening at the appendix and base of the caecum. Colonoscopy showed a sessile polypoid growth at appendicular orifice, at the base of the caecum. Laparoscopy confirmed the clinical suspicion of appendicular carcinoma and laparoscopy assisted radical right hemicolectomy was performed. Final histopathology revealed well differentiated adenocarcinoma of the appendix with no lymph node involvement (pT3N0M0).

Discussion: Patients with primary adenocarcinoma of the appendix present with features similar to acute appendicitis whereas anaemia or fresh bleeding per rectum is a rare presentation. Surgery is the mainstay of treatment, the extent of which will depend upon the stage. Tumours staged as T1 may be managed by appendicectomy alone provided the base is free and there are no lymphadenopathies. T2 or above require right hemicolectomy as chances of lymph node metastasis are high. Nodal involvement warrants the need for adjuvant chemotherapy. Distant metastasis to the peritoneum or liver and lungs is very rare.

Conclusion: While investigating unexplained anaemia or bleeding per rectum, full colonoscopic examination up to the appendicular orifice is important and if required, should be combined with CT scan of abdomen, to clinch the rare but possible and potentially curable diagnosis of appendicular carcinoma.

1. Introduction

Malignant neoplasms of the appendix are extremely rare, with an age-adjusted estimated incidence of 0.12 per 1,000,000 person years [1]. According to the National Cancer Institute, based on data from the Surveillance, Epidemiology, and End Results (SEER), appendicular cancer accounts for 0.4% of all gastrointestinal tumours [2]. There are no established risk factors for the development of appendicular cancer. Malignant appendicular tumours most often present with features similar to acute appendicitis and are diagnosed incidentally at histopathological assessment of the surgical specimen. Appendicular cancers may also be asymptomatic and be found incidentally as an abnormal appearing appendiceal orifice on colonoscopy, at surgery, or on cross-sectional imaging for other indications, such as presumed ovarian

malignancy. In the literature, colorectal polyps are associated with a higher risk of appendicular adenocarcinoma for example with a caecal or rarely a rectal polyp [3,4]. We herein present a case of adenocarcinoma of appendix involving the base of caecum in a background of previously excised dysplastic rectal polyp in an old male patient presenting at a tertiary cancer care centre. This case has been reported in line with the SCARE Criteria [5].

2. Case summary

An 81 year old gentleman of Indian ethnicity with multiple comorbidities including type-2 diabetes mellitus, hypertension and hypothyroidism had presented to our hospital with complaints of fresh bleeding per rectum. He had a history of rectal polypectomy by

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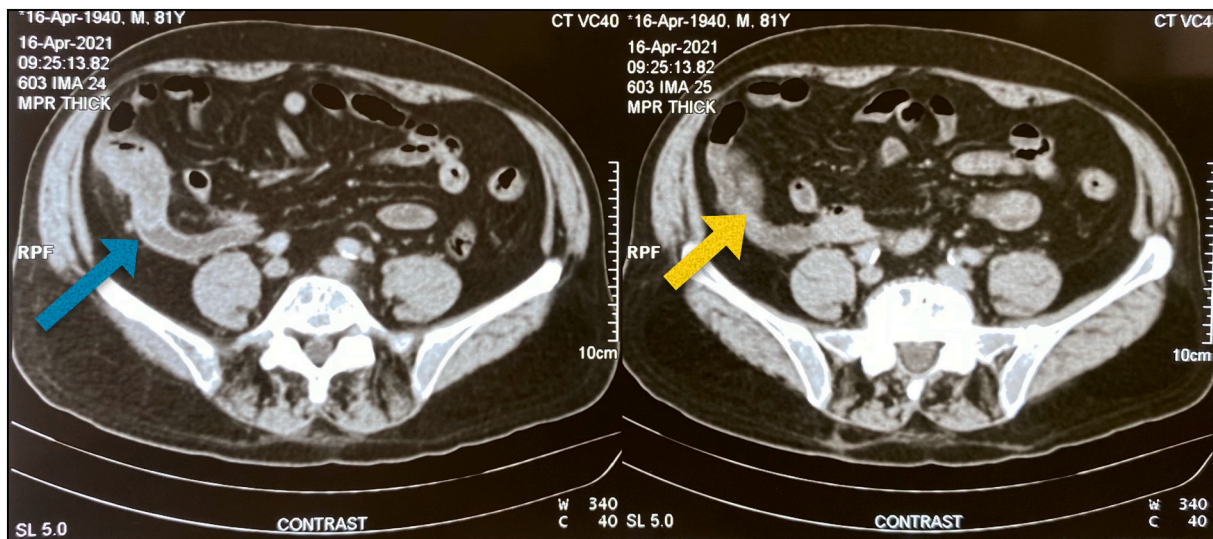


Fig. 1. Axial CECT scan of whole abdomen showing mildly enhanced thickening of the wall of the elongated appendix and the base of the caecum. Arrows indicate thickening and enhancement.

colonoscopic snaring 7 years back which was reported as adenocarcinoma in-situ. On examination, patient had pallor with performance score = ECOG 1. His other general and systemic findings were normal. His haemoglobin level was low (7.2 g/dL) for which he was transfused 2 units of packed red blood cells. Upper GI endoscopy (to rule out any upper GI pathology and evaluate the cause of anaemia) was found to be normal. Colonoscopy showed a few small sessile hyperplastic polyps in upper rectum and single sessile polypoid growth in caecum near the appendicular orifice. Colonoscopic biopsy of the lesion in the caecum showed adenomatous polyp with high grade dysplasia. CECT scan of whole abdomen revealed a mildly enhanced thickening in the wall of the elongated appendix and the base of the caecum (Fig. 1). CT scan of chest was found to be normal.

Multi-disciplinary tumour board evaluation was done and patient was then planned for surgery. Staging laparoscopy followed by laparoscopy assisted right radical hemicolectomy with D2 lymphadenectomy and ileo-transverse anastomosis were performed (Fig. 2). The specimen was delivered outside via a small mid-line incision through a sterile plastic sleeve. The transverse colon was divided at the junction of medial two third and lateral one third of the transverse colon and the terminal ileum was divided at 15 cm proximal to the ileo-caecal valve. Extra-corporeal single layer end-to-end ileo-transverse anastomosis was performed. Operative findings showed a mass involving the whole of appendix extending onto the base of the caecum (Figs. 3 & 4). Final histopathology was reported as well differentiated adenocarcinoma of the appendix extending up to the sub-serosa and base of the caecum with free margins. All 13 harvested lymph nodes were free from metastases (Grade 1-PT3N0M0). Lympho-vascular and perineural invasions were absent (Fig. 5). Immuno-histochemistry (IHC) panel was negative for CDX-2 and Chromogranin-A. Post operative period was uneventful and bowels moved on post-operative day (POD) 3, and patient was discharged on POD 6. A month later, patient was asymptomatic when presented for follow-up.

3. Discussion

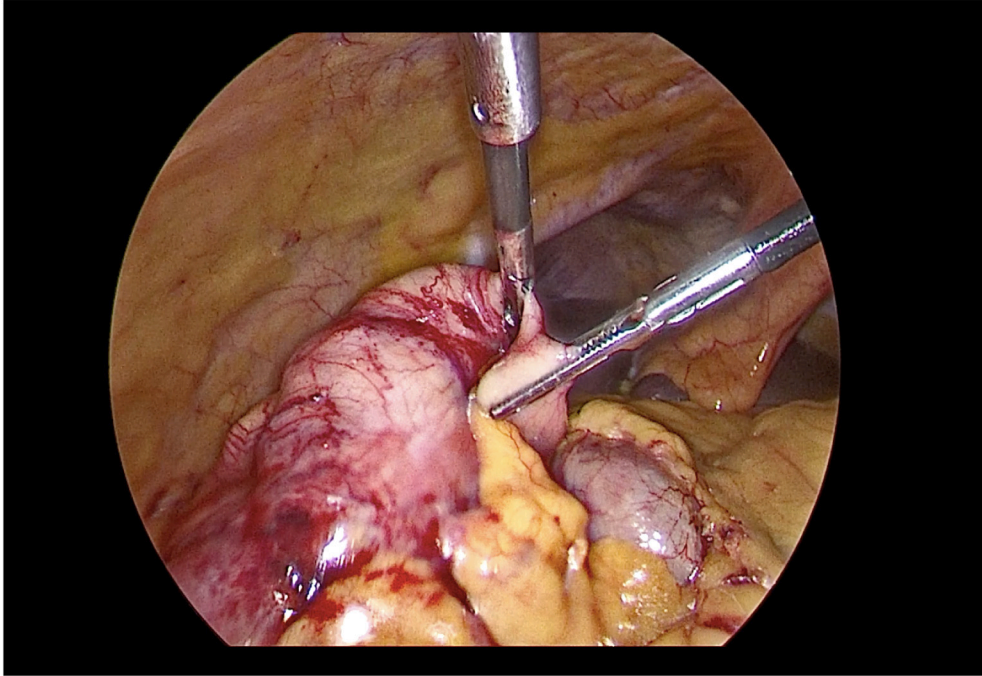
Malignancies of the appendix are extremely rare constituting less than 0.5% of all gastrointestinal tract neoplasms [1,6]. The most common type of appendicular tumours based on their incidences is classified as neuro-endocrine tumours (NET) followed by adenocarcinoma and GISTs. Adenocarcinoma of the appendix maybe further sub-divided into colonic and mucinous signet-ring cell type which are both biologically

and histologically different [7,8]. Similar to colon cancer, colonic-type adenocarcinoma of the appendix presents at a mean age of 62 to 65 years [9,10].

Patients with colonic-type adenocarcinoma of the appendix most often present with incidentally identified lesions following appendectomy for appendicitis or other indication, and will have pathologic T-stage information immediately available. Presentations with bleeding per rectum or unexplained anaemia are very rare [11]. T-is tumours resected with negative margins can be managed with appendectomy alone. T1 tumours with favourable characteristics are thought of like malignant polyps. Appendectomy alone may be sufficient if these lesions are grade 1 or 2, have no angio-lymphatic invasion, and have negative resection margins. All patients should undergo complete colonoscopy to evaluate synchronous colorectal lesions. Although the workup, staging, and treatment of colonic-type adenocarcinoma arising in the appendix mirror that of colon cancer, in the 7th edition of the AJCC Staging Manual, appendiceal carcinomas are classified separately from colorectal carcinomas [12]. Patients found to have un-favourable T1 tumours (high-grade, angio-lymphatic invasion with or without positive margins) should be considered for formal right hemicolectomy for adequate staging and resection. Patients with T2 or greater tumours require complete staging with contrast-enhanced computed tomography (CT) of the chest, abdomen, and pelvis if not already performed. If there is no evidence of distant metastasis, right hemicolectomy is recommended, with 12 or more lymph nodes typically considered adequate for accurate staging. In our case, on imaging and surgical exploration, the tumour appeared to be greater than T2. Hence decision of a formal right hemicolectomy was taken.

In the largest population-based study of primary appendicular cancer, the rate of lymph node involvement in the colonic subtype of adenocarcinoma was 30% [1]. As per the NCCN guidelines, patients with node-positive disease (stage III) warrant adjuvant systemic chemotherapy with 5-Fluorouracil and Leucovorin or Capecitabine with Oxaliplatin if medically fit [13]. As for colonic primary adenocarcinoma, adjuvant chemotherapy should also be considered for stage II patients with high-risk features, especially younger patients and those with inadequate nodal staging [14]. Several factors have been ascertained with regard to better prognosis and lower risks. These include younger age (<50 years old), lower TNM stage (no serosal invasion or distant metastasis), >12 resected lymph nodes without metastasis and well differentiated histological grade. Studies have shown that well-differentiated adenocarcinomas of the appendix carries a better

A



B

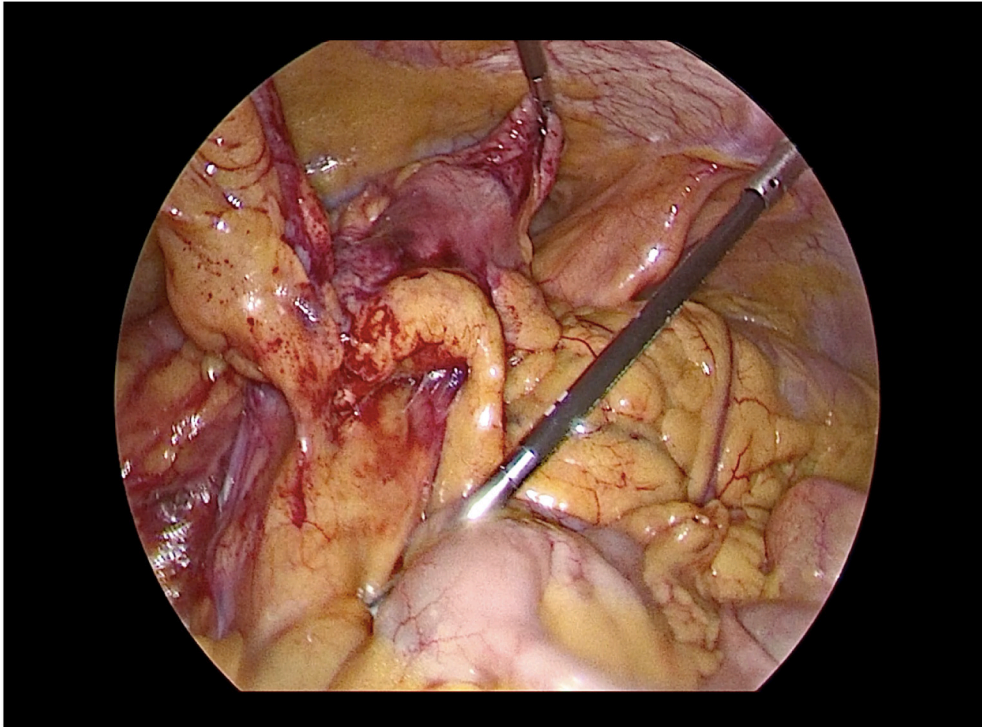


Fig. 2. (A, B) Figure showing laparoscopy assisted right radical hemicolectomy with ileo-transverse anastomosis.



Fig. 3. Appendicular tumour mass involving the base of the caecum delivered through the midline wound with a plastic sleeve.

prognosis than poorly differentiated adenocarcinomas, and histological grade, as defined by the AJCC TNM system, may serve as an important predictor of prognosis for appendicular adenocarcinoma [15]. The final histopathology report in our case ruled out any regional lymph node involvement or any high risk features and therefore adjuvant chemotherapy was not required.

The incidence of distant metastasis at presentation for colonic-type adenocarcinoma of the appendix is reported as 23 to 37% [1,7]. The peritoneum (including ovaries) is the most common site of metastasis followed by liver and lung metastases. Systemic chemotherapy is the recommended treatment for asymptomatic patients who present with distant metastasis. For patients with peritoneal-only metastatic disease, complete cytoreductive surgery and hyperthermic intraperitoneal chemotherapy (HIPEC) should be considered if complete tumour debulking can be achieved [16]. As is true for colorectal cancer, surgical resection, including metastasectomy of limited liver or lung lesions, is

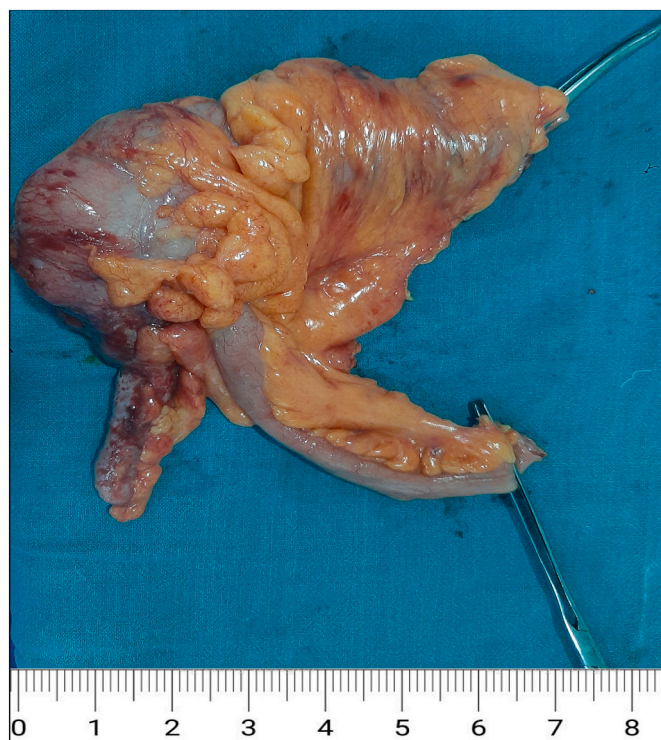


Fig. 4. Post operative specimen of right hemicolectomy with appendicular tumour.

reasonable for select patients with appendix adenocarcinoma.

4. Conclusion

Appendicular carcinoma is rare and mostly diagnosed as an incidental finding after appendicectomy. Elderly people presenting with symptoms of acute appendicitis should raise the suspicion of appendicular malignancy. While investigating unexplained anaemia or bleeding per rectum, full colonoscopic examination up to the appendicular orifice is important. This should be combined with a contrast enhanced CT scan of abdomen, which helps to clinch the rare but possible and potentially curable diagnosis of appendicular carcinoma by a showing a mass in the appendix with enhancement with or without the presence of lymph nodes. Surgery is the mainstay of treatment, the extent of which will depend upon the stage. Adjuvant chemotherapy

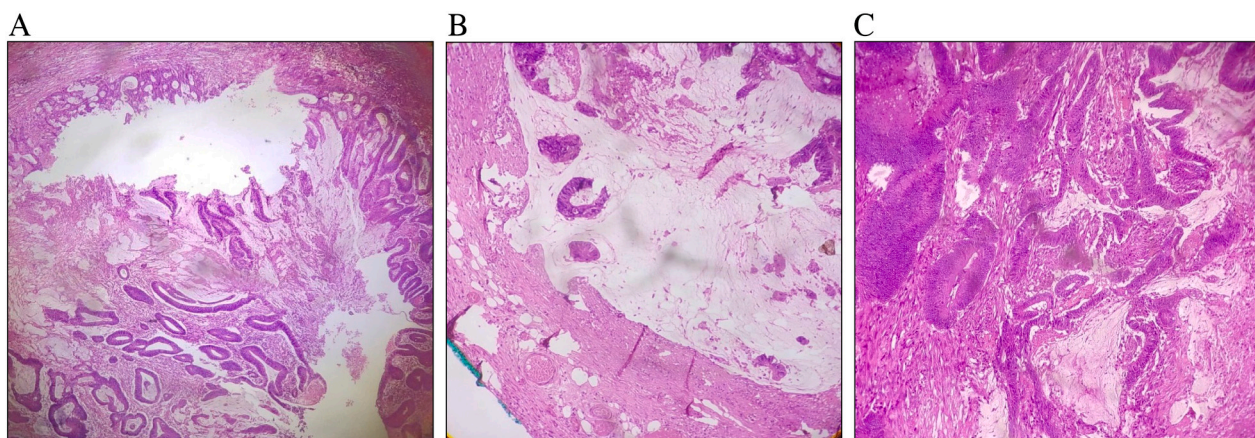


Fig. 5. (A, B, C) Final post operative histopathology slides showing well differentiated Adenocarcinoma of the appendix extending up to the sub serosa with free margins and no regional lymph node involvement (Grade 1-PT3N0M0) on High Power View (40x) in H&E stain.

may be required based on the histopathology report.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Informed consent from patient

Written informed consent was obtained from the patient for publication of this case report along with clinical images without disclosing his identity. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

Exempted from Institutional Ethics Committee of Saroj Gupta Cancer Centre and Research Institute, Kolkata, India.

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Declaration of competing interest

None.

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