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Case report Appendiceal carcinoma presenting as a rapidly enlarging abdominal mass

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|---|---|--|--|--|
| A R T I C L E I N F O Keywords: Appendiceal carcinoma Abdominal mass Rapidly enlarging Right hemicolectomy Adenocarcinoma Appendix | Introduction: Appendiceal adenocarcinomas, although rare, usually present as slow-growing masses. Rarely, do they rapidly enlarge into large abdominal masses over a short period of time. Generally, we attribute rapidly growing abdominal masses to sarcomas, desmoid tumors, or gynecological masses. We present a case of a rapidly growing appendiceal adenocarcinoma in an elderly patient. <i>Case presentation:</i> We report a case of an 83-year-old-male who presented with a one-month history of abdominal pain. A computer tomography (CT) scan identified a lower pelvic mass measuring 7.39 cm × 5.40 cm. A biopsy was done which revealed appendiceal carcinoma. A plan f or radiotherapy to reduce the tumor size was made and the patient was discharged. However, the patient returned after three weeks due to worsening abdominal pain and a CT scan identified a significant enhancement in the tumor size. The patient underwent debulking surgery. Pathology of the specimen identified adenocarcinoma of the appendix. <i>Discussion:</i> A rapidly growing carcinoma of the appendix is rare as they are known to have a slow growth rate. A swift diagnosis and intervention are required as these rapidly enhancing neoplasms in the abdomen can become unresectable and metastasize. Although there are various methods of treatment for appendiceal carcinomas, the rapid growth rate and lack of response to radiation required debulking surgery. <i>Conclusion:</i> There exist a myriad of differential diagnoses for a rapidly growing mass in the abdomen. We are presenting the first documented case of a rapidly growing appendiceal adenocarcinoma. Surgeons should be aware of the variety of differentials in such cases. | | | |

1. Introduction

Adenocarcinomas are derived from the glandular cells of the mucosa with the metastatic property. Given the glandular origin of adenocarcinoma, tumors may develop in the colon, esophagus, lung, pancreas, or prostate. These tumors are usually thought of as slow-growing lesions and are often found incidentally on final surgical pathology. Rarely, are these found to grow rapidly over the course of a few weeks. Diagnosing these rapid growing adenocarcinoma lesions before it becomes symptomatic is often difficult. This case of rapidly growing appendiceal adenocarcinoma has stimulated our interest, at our community hospital, to present this case to increase awareness of such rapidly enhancing lesions which have not been reported in literature before. This report is in line with the Scare Checklist [1].

2. Case

An 83-year-old male, with no significant past medical or surgical history, was admitted to the emergency department at our institution presenting with dull, achy abdominal pain. He complained of right lower quadrant (RLQ) abdominal pain which began a month ago and had gradually worsened and radiated to the rest of the abdomen over time. He claimed to have lost about ten pounds in about two months. Due to the covid-19 pandemic, the patient was reluctant to seek help, until the pain had become unbearable.

A computed tomography (CT) scan was done and a lower pelvic mass of size 7.39 cm \times 5.40 cm was discovered (Fig. 1a). The patient underwent embolization of the feeding vessels to the tumor, in an effort to slow the growth. Colonoscopy conveyed a mass effect at the area of the ileocecal valve. An interventional radiology guided biopsy was done

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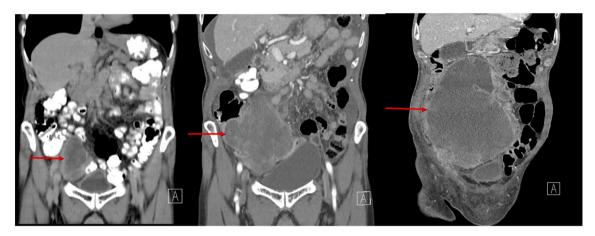


Fig. 1. Coronal CT scan of the abdomen over six weeks prior to the surgery.



Fig. 2. Axial CT scan of the abdomen over 6 weeks prior to the surgery.

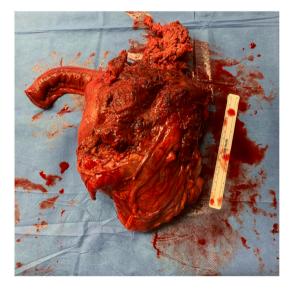


Fig. 3. Gross specimen: en bloc resection of an abdominal mass, right colon, small bowel, and involved peritoneum.

with preliminary findings of adenocarcinoma. The patient was discharged with plans for radiation therapy to shrink the tumor size. However, our patient returned to the emergency room in 3 weeks with worsening pain, and difficulty urinating. A subsequent CT scan showed that the mass (Fig. 1b) size increased to 13.84 cm \times 11.33 cm in three weeks. The patient was unable to receive radiation treatment due to the pain, rapidly enlarging tumor size, abdominal girth, and total encasement of the abdomen. The case was discussed at a multidisciplinary conference, and a decision was made to offer the patient a debulking

surgery. A final CT scan before the surgery measured the mass to be 18.75 cm \times 14 cm (Fig. 1c).

Upon surgical exploration, a large abdominal mass encroached on the midline. The cystic fluid was encountered and aspirated and sent for fluid cytology. The mass stemmed from the right lower quadrant and encased the terminal ileum inferiorly, tracked into the ascending colon, and peritoneum anteriorly. En-bloc resection was performed which included a formal right hemicolectomy, partial resection of the anterior peritoneum. Surgical clips were placed at the margins for future

Table 1

Comparison of different neoplasms.

| Neoplasm | Origin & location | 5-year survival+ | Metastatic at presentation | Management |
|----------------|---|---------------------|----------------------------|--------------------------------------|
| Adenocarcinoma | Mucus producing glandular cells; colorectal, breast, lung, pancreas, prostate, appendix | ~61% [5] | 23–37% [6] | Hemicolectomy [6] |
| Angiosarcoma | Endothelial derived soft tissue tumors; head, neck, breast, small bowel | ~35% [7] | 16-44% [8] | Surgery, chemotherapy, radiation [9] |
| Desmoid tumors | Monoclonal fibroblastic proliferation from deep soft tissues; abdomen, arm, leg | ~95% [10] | Non-metastatic [11] | Surgical resection [12] |
| Leiomyosarcoma | Mesenchymal origin, smooth muscle soft tissue; abdomen, uterus | ~45% [13] | 63% [14] | Surgical resection [15] |

radiation treatment. No evidence of grossly metastatic disease was apparent on the liver or the remaining mesentery and omentum. The abdomen was closed primarily. The patient had an uneventful recovery and was discharged home on postoperative day 3 with follow-up for radiation treatment. The pathology report identified Cam 5.2, CDX2, AE1/AE3 tumor markers which pointed to a diagnosis of adenocarcinoma of the appendix (Figs. 2 and 3).

3. Discussion

Rapidly growing abdominal masses in adults are rare, and their diagnosis is associated with high morbidity and mortality [2]. Given that the majority of abdominal tumors are non-palpable upon presentation, clinical diagnosis becomes the primary method of diagnosis [3]. The top differential for such a rapidly growing mass is usually a sarcoma. These tumors may rapidly become unresectable when locally advanced or when metastasis occurs. This raises concern as the majority of the GI tract tumors are slow-growing. Therefore, early diagnosis and treatment are associated with improved outcomes. Diagnosis is often due to persistent gastrointestinal symptoms or incidental detection on imaging. CT scan findings are confirmed by tissue diagnosis following operative resection. In this case, three CT scans were done over a period of 6 weeks. The lesion had a very high rate of enhancement over this interval which is quite rare. Appendiceal masses are usually slow-growing masses [4].

When possible, surgery is the ultimate treatment for these tumors. R0 Resection of appendiceal adenocarcinoma is associated with 61% 5-year survival (Table 1). [5] Unresectable tumors have a poor prognosis. Local tumor burden often mandates surgical treatment with curative or palliative intent. This differentiation is usually done intra-operatively. Treatment is based on expert opinion and surgeon preference. While the primary management for appendiceal cancers is right hemicolectomy; radiation and chemotherapy are also treatment options for appendiceal adenocarcinoma. The severe pain, lack of response to radiation, and rapid growth of the mass, in this case, necessitated a debulking surgery. We performed an en-bloc resection of the terminal ileum, right colon, and the involved peritoneum. The surgeon should remember to place clips at the margins to aid with targeted radiation therapy, when such large masses are encountered.

A CT scan is the optimal diagnostic modality in a patient with an abdominal mass. Serial CT scans should be done to monitor the size of the mass across different diagnoses. The current NCCN guidelines for follow-up on patients with abdominal masses with biopsy-proven malignancy is serial CT of intervals varying from 3 to 12 months depending on the stage of the tumor and serial CEA markers.

While rapid growth despite active treatment is not typical of abdominal masses, this patient is not unique. A neoplasm like an adenocarcinoma is often considered to be a slow-growing neoplasm [4]. There have been three other cases reported with rapidly growing abdominal adenocarcinoma that should also be considered [16]. Although rare, these occurrences should remind us that rapidly growing lesions do exist and increasing awareness of such rapidly expanding lesions may be crucial for rapid treatment and diagnosis. This report serves as an instrument to increase awareness and point out a possible treatment plan for such lesions.

4. Conclusion

Appendiceal adenocarcinomas, although rare, usually present as slow-growing masses which are typically found after an appendectomy. The top differentials for a rapidly growing abdominal mass usually include sarcomas, desmoid tumors as well as gynecologic etiologies. We describe the first documented case of an appendiceal adenocarcinoma presenting as a large, rapidly expanding intra-abdominal mass. Surgeons should be aware of the wide array of differentials that may exist in the presence of abdominal masses that are growing rapidly.

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Ethical approval

N/A.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration (for case reports detailing a new surgical technique or new equipment/technology)

N/A.

Guarantor

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Declaration of competing interest

None.

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