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Case Report

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PRIMARY ADENOCARCINOMA OF THE APPENDIX: A RARE ENTITY

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ABSTRACT

Primary adenocarcinoma of the appendix is a rare entity. It is often diagnosed first on histopathology in specimen submitted for suspected appendicitis. Neoplasms of the appendix are found in around 1% of appendicectomy specimen, the frequency of primary appendiceal carcinoma being as low as 0.05-0.2% of the specimen investigated. We report the case of a 60 year old male who presented with acute pain in lower abdomen. Ultrasonography demonstrated appendiceal inflammation and the patient underwent laparoscopic appendicectomy. Histopathological examination revealed adenocarcinoma of the appendix. A retrospective analysis was performed to look for a primary site other than appendix. Thorough examination did not reveal any other site in body harbouring a malignancy and it was confirmed as a primary adenocarcinoma of the vermiform appendix. Thus, histopathology is of utmost importance in all appendicectomy specimen to rule out malignant pathology.

KEYWORDS: Primary adenocarcinoma, appendix.

INTRODUCTION

Primary adenocarcinomas of the appendix are very rare neoplasms accounting for 0.05%- 0.2% of all appendicectomies.[1] They are frequently discovered by the pathologist following appendicectomy for suspected appendicitis.[2] The majority of appendiceal cancers are carcinoids, while others constitute mucinous cystadenocarcinomas, adenocarcinomas, lymphosarcomas, paragangliomas and granular cell tumors. Majority of appendiceal neoplasms present with symptoms of acute appendicitis, i.e., fever, leukocytosis and acute abdominal pain. Appendicectomy is often performed when clinically indicated.[3] Carcinomas of the appendix are usually well differentiated adenocarcinomas, which tend to produce pseudomyxoma peritonei and do not metastasize until late in the disease process.[4]

CASE REPORT

A 60 year old male presented with acute abdominal pain of 2 days duration, along with vomiting and low grade fever. There was no history of loss of appetite, weight loss or any alteration in bowel and bladder habits. Also, there was no history of similar episodes in the past. The patient had a past history of blunt trauma abdomen, for which he underwent an abdominal surgery 25 years back. Systemic examination was within normal limits except for tenderness in the right iliac fossa. All routine investigations were within normal limits. Ultrasonography was done to ascertain the cause of abdominal pain and it revealed appendiceal inflammation along with incisional hernia at the site of previous surgery. The patient underwent laparoscopic appendicectomy and was discharged after 2 days of observation. The appendicectomy specimen measuring 7x4x2 cm was received in our department. On gross examination, the appendix was soft in consistency and appeared congested. Cut section showed dilated lumen filled with small amount of mucin. Histopathological examination of the specimen surprisingly revealed adenocarcinoma of the appendix, characterised by irregular glands infiltrating the entire thickness of the wall with intraluminal mucin extravasation. Glands were lined by pseudostratified columnar cells having large pleomorphic vesicular nuclei along with inflammatory infiltrate and areas of necrosis (Figures 1 and 2).

A retrospective analysis was performed to look for a primary site other than appendix. Thorough examination did not reveal any other site in body harbouring a malignancy. Thus, it was confirmed as a primary adenocarcinoma of the vermiform appendix, colonic type.

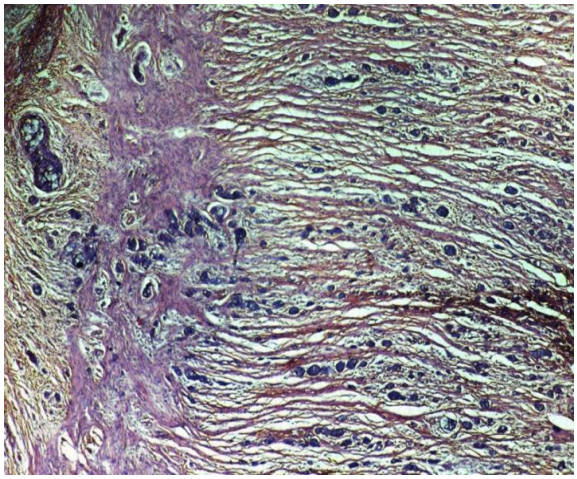

FIGURE 1: Photomicrograph showing irregular glands infiltrating the entire thickness of the wall with intraluminal mucin extravasation (H&E, 100x)

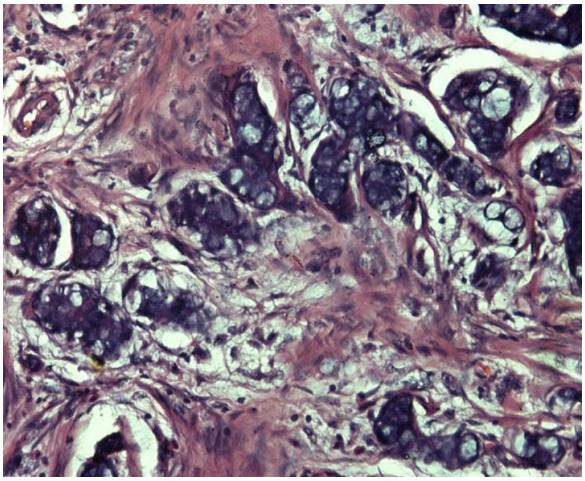

FIGURE 2: Photomicrograph showing irregular glands lined by columnar cells having large pleomorphic vesicular nuclei with inflammatory infiltrate and areas of necrosis (H&E, 400x).

DISCUSSION

Primary adenocarcinomas of the appendix are extremely rare and are rarely diagnosed preoperatively. They most often present as suspected acute appendicitis, probably due to early malignant obstruction of the appendiceal lumen leading to superimposed infection, or as a palpable abdominal mass.[5] Some may even be entirely asymptomatic. The mean age at diagnosis is in the fifth decade of life, with an even male to female ratio for all except for the colonic type, which may have a slight male preponderance.[3] Carcinomas of appendix are usually classified into three distinct subtypes with varying frequencies: mucinous type (55%), colonic type (34%) and adenocarcinoid (11%) which has a mixed morphology. Mucinous and non-mucinous carcinomas of the appendix have similar genetic alterations but different clinical presentation and prognosis. Mucinous histology has a better prognosis than colonic histology.[5]

Computed tomography is helpful in identifying a possible appendiceal neoplasm. Calcification may indicate the presence of a neoplastic process rather than infective or inflammatory. There may be some air foci noted on the CT in cases of associated superinfection.[3] If a patient presents with a distended abdomen due to pseudomyxoma peritonei, a CT would show widespread heterogenous locules in peritoneal cavity. Since there are no imaging studies specific for diagnosing adenocarcinomas, the final diagnosis is most often made postoperatively on histopathological examination.[6]

Right hemicolectomy is the preferred surgical intervention for all subtypes of adenocarcinoma since most tumors present as advanced invasive carcinomas. Some surgeons suggest that a simple appendectomy is sufficient for pT1 carcinoma of the appendix.[2] Role of adjuvant chemotherapy for adenocarcinoma of the appendix is yet to be established. Some studies suggest a response to regimens containing fluorouracil. Malignant involvement of the locoregional lymph nodes is seen in about one third of patients, especially in advanced invasive tumors. Adjuvant chemotherapy is recommended in these patients with lymph node positivity or locally advanced tumors in accordance with the recommendation for colon carcinomas.[6] Quite frequently, carcinomas of appendix show local infiltration in the surrounding peritoneum with increased risk of locoregional recurrence. For these patients with peritoneal tumor metastasis, cytoreductive surgery in combination with hyperthermic chemotherapy has been suggested as a therapeutic option.[7]

Studies investigating the biological alterations of appendiceal carcinomas have reported classic mutations associated with sporadic colorectal cancer including frequent KRAS mutations, rarely p53 overexpression, with majority of tumors being microsatellite stable. Some reports also mention appendix cancers with high grade microsatellite instability, which seems to represent a distinct tumor etiology and might describe an unidentified alteration in the DNA repair pathway.[2]

Prognosis of adenocarcinoma depends on the subtype and extent of disease. Mucinous adenocarcinoma has a more favorable prognosis because it does not exhibit hematogenous or lymphatic spread. Patients with intraperitoneal seeding have worse prognosis.[6] Many studies have also shown that there is a clear survival benefit to the addition of a hemicolectomy. In a study by Nilecki et al, the 5 year survival rate for hemicolectomy was 73% versus 44% in the appendicectomy group.[8]

CONCLUSION

Cancers of the appendix represent a challenge for diagnosis and management. No standard of care has been established so far due to rarity of occurrence. They may

present as acute appendicitis and often operated. Histopathology is of utmost importance in all appendicectomy specimen to rule out malignancy.

REFERENCES

1. Behera PK, Rath PK, Panda R, Satpathi S, Behera R. Primary appendiceal mucinous adenocarcinoma. Ind J Surg, 2011; 73(2): 146-8.
2. Komm M, Fesl MK, Kremer M, et al. Primary mucinous adenocarcinoma of the vermiform appendix with high grade microsatellite instability. J Cancer, 2011; 2: 302-6.
3. Ruoff C, Hanna L, Zhi W, Shahzad G, Gotlieb V, Saif MW. Cancers of the appendix: review of the literatures. ISRN oncology, 2011 Aug 11; 2011.
4. Carr NJ, Emery TS, Sobin LH. Epithelial neoplasm of the appendix and colorectum. Arch Pathol Lab Med, 2002; 126(7): 837-41.
5. McCusker ME, Coti TR, Clag LX, Sobin LH. Primary malignant neoplasm of the appendix. Cancer, 2002; 3307-12.
6. Wanf HL, Lin CK, Lin CC. An appendix adenocarcinoma mimicking appendicitis. Advances in Digestive Medicine, 2016; 3: 73-5.
7. Marcotte E, Sideris L, Drolet P, et al. Hyperthermic intraperitoneal chemotherapy with oxaliplatin for peritoneal carcinomatosis arising from appendix: preliminary results of survival analysis. Ann Surg Oncol, 2008; 15: 2701-8.
8. Nilecki SS, Wolff BG, Schlinkert R, Sarr MG. The natural history of surgically treated primary adenocarcinoma of the appendix. Ann Surg, 1994; 219: 51-7.