



Primary Adenocarcinoma of Appendix Presenting As Acute Appendicitis— A Case Report

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ABSTRACT

It is extremely rare for mucinous adenocarcinoma to develop as a primary tumour. The incidence of epithelial malignancies of the appendix has been estimated to be 0.12 per 1 million persons per year. We report a case of primary appendiceal adenocarcinoma of signet ring cell variant presenting with features of acute appendicitis. Routine investigations showed dilated appendix with peri-appendiceal fluid collection suggesting acute appendicitis. After appendicectomy, the specimen was sent for routine histopathological examination. The diagnosis came out to be signet ring cell adenocarcinoma. As a second stage procedure, right hemicolectomy was done. As it was an incidental finding, this case emphasizes that routine histopathological examinations of all appendicectomy specimens is essential and informative.

Keywords- Appendicitis, histopathology, mucinous, adenocarcinoma. Signet ring cell adenocarcinoma.

INTRODUCTION

Primary adenocarcinoma of the appendix are classified into 4 major groups-mucinous adenocarcinoma, colonic type adenocarcinoma, goblet cell carcinoma, and signet ring cell carcinoma. ^[1] Of all these, signet ring cell type adenocarcinoma is extremely rare and colonic type is most common. Carcinoma of the appendix is usually well differentiated mucinous adenocarcinoma, which tend to produce pseudo-myxoma peritonei and do

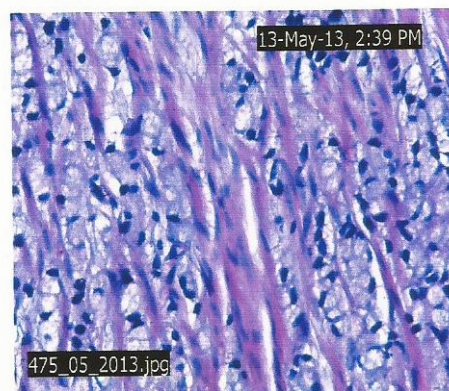
not show metastatic spread until late in the disease pathway.^[2] Primary appendiceal adenocarcinoma are very rare malignant tumours .accounting for 0.05-0.2 of all appendectomies and only 6% of all malignant tumours of the appendix.^[3] We report a rare case of signet ring cell adenocarcinoma of the appendix in a lady of fifty years of the Munda community who initially presented as pain abdomen and features of acute appendicitis. We will also briefly review the topic of appendiceal carcinoma.

CASE REPORT

A fifty year old woman presented in the emergency room with severe pain abdomen in the right lower abdomen of two day duration, associated with vomiting and low grade fever. There was no history of weight loss, loss of appetite or involvement of bowel and bladder. Review of systems was unremarkable.. Past medical and surgical histories revealed a caesarean section for two children twenty years back. Abdominal examination revealed tenderness in the right iliac fossa. Ultrasonography (USG) revealed a dilated non compressible tubular structure in the right iliac fossa, with peri-appendiceal free fluid. A diagnosis of acute appendicitis was suggested. Laboratory findings revealed leucocytosis. Complete blood count, coagulation profile, basic metabolic panel was unremarkable. Emergency appendicectomy was done . Intra-operative findings showed a dilated, inflamed appendix with little fluid collection around it. No mucinous material was found inside the abdominal cavity. The specimen was sent for histopathological examination. On gross examination, the appendix was enlarged to 8 cmx1 cm in size, firm in consistency with tortuous veins on its surface Cut section showed wall of appendix transmurally infiltrated by a tumour upto the periappendicular adipose tissue and composed of sheets of cells with eccentric hyperchromatic nuclei and abundant cytoplasm. The impression was signet ring cell adenocarcinoma of the appendix. Right hemicolectomy was done as a second stage procedure.

DISCUSSION

Adenocarcinoma of the appendix is rare. In 12-15 % of cases, a second malignancy in the gastrointestinal tract is associated with the disease ^[4]. Adenocarcinoma of the appendix is estimated at around 0.2/100000 per year. Tumours of the appendix are found in approximately 1% of appendiceal specimens submitted for pathological examination. The American national cancer Institute's Surveillance, Epidemiology and End results (Seer) program analysed 1,645 appendiceal malignancy cases from 1973—1998. 37.2% of the cases were identified as muinous adenocarcinoma, 24.9% "colonic type", 19.6% "malignant carcinoid", 13.7% "goblet carcinoid" , and 4.3% "signet ring cell" carcinoma.^[5] Connor et al. reviewed a database of 7970 appendectomies and found 74 patients with



Slide of *Signet ring cell adenocarcinoma*. appendiceal tumours: 42 carcinoid, 12 benign, and 20 malignant^[9] Adenocarcinoma of the appendix most often manifest as acute appendicitis or as a palpable abdominal mass. But some are entirely asymptomatic.^[8] Malignancies of the appendix are never suspected preoperatively and seldom intraoperatively, the diagnosis being made at histopathological examination of the surgical

specimen or as an incidental finding during exploration for another disease. Although ultrasonography (USG) can be used to evaluate an abdominal mass, CT is superior to USG in regards to anatomical topography of an appendiceal adenocarcinoma with the ability to distinguish between caecum and mucocoele, as well as the ability to detect mural calcifications within the neoplasm. An appendiceal diameter greater than 15 mm is not specific, but this finding should be viewed with extreme suspicion of appendiceal malignancy.^[10] The optimal treatment of any adenocarcinoma of the appendix is right hemicolectomy, either as a primary operation or as a secondary operation after adenocarcinoma of the appendix is noted on microscopical examination.^[11] When appendiceal cells pass into the peritoneal cavity and incite pseudomyxoma peritonei, a catastrophic complication develops. Care must be taken regardless of the approach when handling this neoplasm. Patients with appendiceal adenocarcinomas have a significant risk of synchronous and metachronous neoplasm, which often originate from the gastrointestinal tract. Staging for appendiceal adenocarcinoma uses the tumour –node- metastasis classification similar to colonic adenocarcinoma^[12]. Patients with low grade neoplasms confined to the appendix had a 100% survival at a median survival time of 6 years.^[13] Those with extra-appendiceal spread had a survival at 86% at 5 years.^[14] Right hemicolectomy with appropriate adjuvant therapy is recommended for both mucinous and non-mucinous carcinomas.^[15] The prognosis of mucinous adenocarcinoma depends on the grade of malignancy of the

mucinous tumour and success of debulking surgery depends on the extent of removal of the tumour that have metastasized to the abdomen^[16].

CONCLUSION

Signet ring cell adenocarcinoma of the appendix is a rare entity that in most cases is found incidentally. We would like to stress on histopathological examination of the appendectomy specimen on a routine basis, to rule out malignant pathology.. This case is being reported because of its rarity and interesting nature.

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