



Adenocarcinoma of Appendix- A Case Series

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Abstract

Neoplasms of appendix are rare and may be encountered at any elective or emergency operation. It is estimated that 50% of appendicular neoplasms present as appendicitis and are diagnosed on pathologic examination of surgical specimen. The incidence is 0.4% to 1% Of GI malignant neoplasms. Carcinoid tumors are the most common primary tumor identified in the appendix. Adenocarcinoma of the appendix is very rare and occurs at a frequency of 0.08% to 0.1% of all appendectomies. Moreover adenocarcinoma of appendix occurring in pregnancy is rare and may present as an acute appendicitis. Here in this study, we report a series of three cases which are diagnosed as adenocarcinoma of appendix. One of which occurring in pregnancy.

Keywords: Appendix; Adenocarcinoma; Mucin

Introduction

Adenocarcinoma of appendix is a rare disease. It occurs in 0.1% to 0.2 % of appendectomies incidence of disease is 0.2/1,00,000 per year. Only few cases are reported for its occurrence in antenatal mother. The most common age group is 6th or 7th decade of life and it is seen most commonly in males. 75% of cases present with clinical symptoms 25% cases are detected as incidental findings grossly, low stage lesions (tumor confined to appendix wall) present as thickening of appendix. Lesions may spread beyond the appendix wall present as masses. In which 1/4th of cancers are cystadenocarcinomas, present as mucoceles. The most common histologic pattern is fairly well differentiated mucinous adenocarcinoma. Etiology is unclear and there is an association with neoplasia elsewhere in large intestine. Risk factor includes chronic ulcerative colitis in primary adenocarcinoma. This case series has been reported due to its rarity, the paucity in clinical symptoms, its incidence in antenatal mother and the diagnostic dilemma thus was caused by the disease.

Case Presentation

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Case 1

A 42-year old female presented with abdominal pain for one month with abdominal distension and vomiting for 1 week. CECT abdomen was done showing mesenteric cyst of size 15 cm × 10 cm as shown in Figure 1. Diagnosed to be a mesenteric cyst in the right iliac fossa. Exploratory laparotomy was done. Per op finding- a cyst of size 14 cm × 6 cm × 6 cm was present adjacent to the caecum originating from the tip of the appendix as shown in Figure 2. The cyst contained mucus along with pus and necrotic material. Appendectomy was done and specimen sent for HPE. Histopathological examination shows mucin producing adenocarcinoma of the appendix. Medical oncology opinion was obtained and she was started on chemotherapy (gemcitabine and carboplatin) and planned for chemotherapy for Re-staging.

Case 2

An 18-year old antenatal mother 11 weeks of gestation, presented with abdominal pain for 1 week, anorexia and nausea. Diagnosed to have acute appendicitis and started on IV antibiotics and analgesics. After 2 weeks patient developed features of acute appendicitis, proceeded with laparoscopic appendectomy. Per op finding is long inflamed kinked appendix paracaecal in position as shown in Figure 3. Histopathological examination of specimen shows adenocarcinoma of appendix as shown in Figure 4. Immunohistochemistry shows positivity for CK20 infiltrating glands and negative for chromogranin. Patient in regular follow up. And planned for further management and staging after childbirth.

Case 3

A 40 year old male presented with acute abdominal pain and diagnosed to have acute appendicitis. Patient underwent emergency open appendectomy. Gross specimen shows specimen

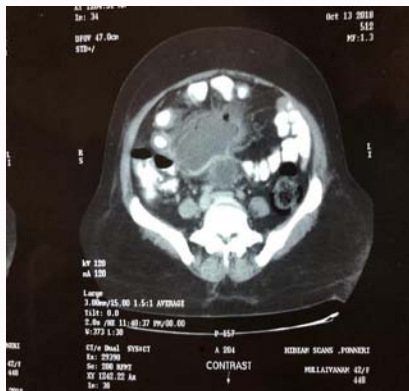


Figure 1: CECT showing mass in abdomen of case report 1.

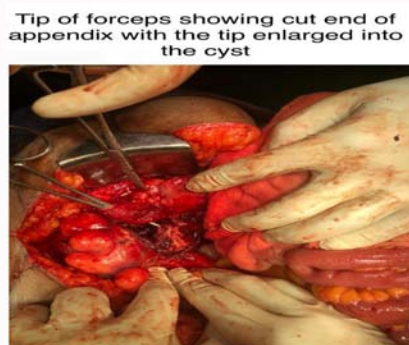


Figure 2: Intraoperative picture of case report 1.

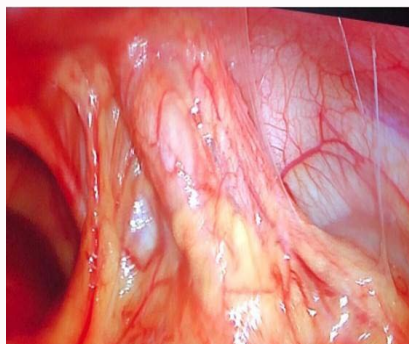


Figure 3: Laparoscopic view of case report 2.

of appendix measuring 7 cm × 2 cm × 2 cm with external surface showing exudative membrane present. Cut Surface shows lumen to be dilated, filled with mucinous material as shown in Figure 5. Patient was discharged as post op period is uneventful. Patient came for regular follow up with HPE report shows Extensive ulceration of lining epithelium with sub-epithelium showing granulation tissue with pools and pockets of mucin as shown in Figure 6. Tumor cells exhibits mild degree of nuclear atypia with Occasional signet ring cells seen. Mucin pools are seen to dissect into muscularis propria.

Serosa shows numerous congested blood vessels. Patient was reported as mucinous adenocarcinoma appendix, well differentiated grade 1. Since base of appendix was involved by tumor, patient was taken up for right hemicolectomy which showed ceecal infiltration; post-operatively patient was started on adjuvant chemotherapy. Patient in regular follow up now for the course of adjuvant

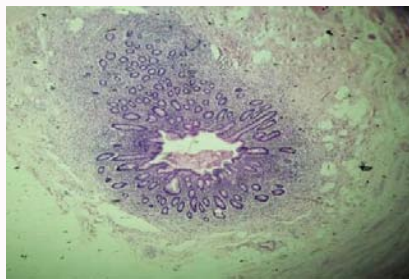


Figure 4: HPE picture of case report 2.



Figure 5: Gross specimen of case report 3.

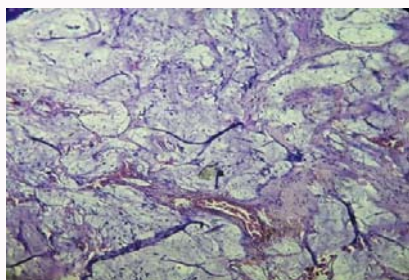


Figure 6: HPE picture of case report 3.

chemotherapy.

Discussion

Primary adenocarcinoma of the appendix is rare disease with less than 300 cases reported in literature. It is typically found in adults of 50 years to 60 years old, they usually presents with symptoms of acute appendicitis. Preoperative detection of appendiceal adenocarcinoma is rarely possible, unless they are large tumors involving the stump of appendix. Adenocarcinomas of the appendix show characteristic morphological variations, from those that resemble the usual colorectal carcinoma, to those that arise from a carcinoid, to those alike mesenteric cyst, to mucinous tumors that may appear well differentiated and indistinguishable from adenoma. Mucinous adenocarcinoma and colonic adenocarcinoma are the most common types that are found in appendix. 40% of them are mucin secreting mucinous adenocarcinomas. Classifying the type of adenocarcinomas is important because colonic adenocarcinoma has a bad prognosis as compared to mucinous adenocarcinoma.

The presence of Metastatic mucinous adenocarcinoma deposits from elsewhere should be examined in order to plan the treatment especially in female patients where mucinous adenocarcinoma of ovary must be ruled out. Appendiceal stump must be evaluated thoroughly because involvement of stump of appendix requires right hemicolectomy. Duke staging has to be done on tumors of appendix,

since stage C1 and C2 require adjuvant chemotherapy. Peritoneal accumulation of gelatinous ascites caused by malignant neoplasms of the vermiform appendix is an important finding that must be looked in all adenocarcinomas of appendix as it alters the staging of the tumor.

The incidence of appendicitis in pregnancy (0.05% to 0.07%) is similar to that in the general population. Acute appendicitis is not rare in the first semester of pregnancy. It accounts for almost 50% of all pregnancy-related appendicitides. Appendicitis is difficult to diagnose in pregnancy: the clinical presentation often varies and diagnosis is usually delayed. Right-side abdominal pain is the principal basis for diagnosis, while leukocytosis and low-level fever, as in the non-pregnant state, are unreliable for diagnosis. Pregnant patients are more likely to present with perforations possibly due to the immunosuppression of the pregnancy (43% vs. 4% to 19% in the general population) [1-5]. Perforations in pregnancy often lead to serious complications, including peritonitis and fetal death, however the risk of perforation appears to increase with gestational age, and thus it is rarer [6-8]. The gynecologists should be aware of the possibility of appendicitis in any pregnant woman that presents with a new or acute abdominal pain. Appendicitis should be diagnosed and appendectomy should be performed as early as possible in patients presenting a highly suggestive clinical and ultrasonography picture, preferably by laparoscopy, in order to avoid more severe complications which could be life-threatening for the mother or the fetus. Adenocarcinoma of the appendix is very rare especially in appendicitis complicating pregnancy and occurs at a frequency of 0.08% to 0.1% of all appendectomies done for appendicitis.

Mucinous adenocarcinoma is the most common cancer of the appendix. It accounts for 37% of all appendix neoplasms. This tumor may grow faster and can metastasize to the lymph nodes, liver and lungs. Survival in this type of tumor group is significantly worse than the other appendiceal carcinomas and it should be considered as a separate type of appendiceal malignancy because of its poor prognosis. The term for extensive spread of these tumors in the abdomen is peritoneal mucinous carcinomatosis. It can also lead to pseudomyxoma peritonea that is commonly used to refer to widespread mucinous disease in the abdomen caused by either mucinous adenoma or mucinous adenocarcinoma. The presentation can mimic acute appendicitis, Right Iliac Fossa (RIF) mass and intestinal obstruction [6]. Mucinous neoplasms can also present with

uncommon anatomical anomalies such as intestinal malrotation and *situs in vs.* right hemicolectomy is considered to be the treatment of choice for the lesion beyond the mucosa and appendectomy alone for a localized lesion.

The role and safety of laparoscopic appendectomy for management of the incidentally discovered appendiceal tumors has not been established. Adjuvant multimodal treatment prevents the disease progression. One of the related studies mentioned that three cycles of Capecitabine 1250/m² from day 1 to 14, oxaliplatin 130 mg/m² on day 1, and (CAPOX) every 21 days, followed by a total dose of 50.4 GY external-beam radiation therapies improved the life. This case series has been reported due to its rarity, the paucity in clinical symptoms, its incidence in antenatal mother and the diagnostic dilemma thus was caused by the disease.

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