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Case Report

Primary signet-ring cell carcinoma of vermiform appendix: a rare case report

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ABSTRACT

Appendiceal primary signet ring cell carcinoma is an extremely rare neoplasm considered to be more aggressive than other appendiceal tumour. Primary appendiceal carcinoma is diagnosed in only 0.9%–1.4% of appendectomy specimens and signet-ring cell carcinoma (SRCC) of vermiform appendix is accounting 0.43% of all appendiceal malignancies. Patients of SRCC usually present clinically as acute appendicitis. Preoperative imaging detection of appendiceal adenocarcinoma have limited benefit due it has minimal change like appendicitis or wall thickening without any obvious growth. We reported an extremely rare case of primary signet ring cell carcinoma of the vermiform appendix in a 55 year old man. Patient had abdominal pain, anorexia and nausea. He underwent appendectomy for appendicitis and histopathologically diagnosed as appendiceal signet ring cell carcinoma with lymph node metastasis.

Keywords: Signet ring cell carcinoma, Appendix, Intestine, SRCC.

INTRODUCTION

Appendiceal cancer is rare cancer; it accounts for only 0.5% of all gastrointestinal neoplasms.[1] According to a nationwide cancer database (SEER), the age-adjusted incidence of appendiceal malignancies was 0.12 cases per 1,000,000 people per year.[1] Primary appendiceal cancer is diagnosed in only 0.9%–1.4% of appendectomy specimens.[2] Further, signet-ring cell carcinoma (SRCC) of vermiform appendix is extremely rare, accounting for 0.43% of all appendiceal malignancies.[2] Our case is an extremely rare case of primary appendiceal SRCC histologically which clinically presenting as acute appendicitis.

CASE REPORT

A 55-year-old Indian man presented with right lower abdominal pain and nausea since 3 months, weight loss 5 kg in 3 months and anorexia since three and half months and physical examination showed abdominal tenderness. He was diagnosed with acute appendicitis by clinically and admitted to our hospital. Ultrasonography demonstrated appendiceal wall thickening (7.2 mm). Rest of intestine and other abdominal organs were normal in ultrasonography. Blood test showed mild leucocytosis (11,200 cumm: normal 4000–11000 cumm). He was further planned for operation. An appendectomy using laparoscopy was performed. Gross examination during the operation showed mildly inflamed appendix. .The size of appendix was 4 cm.

Gross pathological examination showed appendix measuring 4 cm with no apparent tumour, but the whole appendix showed wall thickening and lumen obliterated (Figure 1). After fixation in formalin, the appendix was cut into three sections, and was observed under microscopically.

Microscopy - Multiple sections were taken and stained with haematoxylin and eosin stain. Microscopy showed single and nests of epithelial cells in full thickness of wall of appendix were seen in the two sections except surgical end which was free from cancer cells. Cells were present

in nests, solid sheets, glands and single cells pattern (Figure 2). Cells showed abundant cytoplasm with large single vacuole containing mucin with displacing nuclei at periphery (Figure 3). These cells forming 70% burden of total cancel cells. Other cells had moderate abundant cytoplasm with central to eccentric nuclei. The carcinoma cells invaded into the muscular layer and serosa also. Vascular and neural invasions (Figure 5) were also present. These malignant cells were PAS positive & diastase resistant for intracytoplasmic mucin (Figure 4 & 5). After diagnosis right hemicolectomy was performed. Hemicolectomy specimen along with mesentery had not any growth grossly. Three lymph node grossly identified in which largest measured 1.2*0.4 cm. In microscopy, whole resected segment of colon and mesentery were free from carcinoma cells. Two lymph nodes were positive for SRCC metastasis.

Figure 1: Show specimen of appendix with obliterated luman.

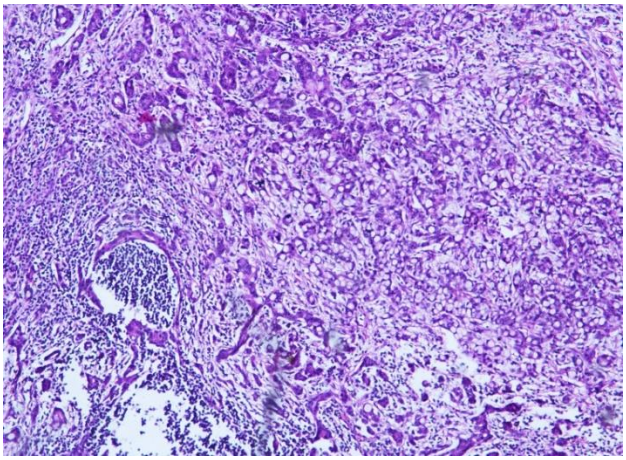

Figure 2: Section shows signet ring cells diffusely infiltrating appendiceal wall (H&E, 10x).

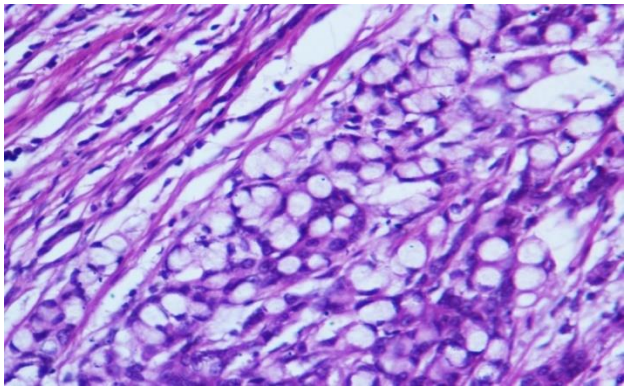

Figure 3: Section shows signet ring cells having large single cytoplasmic vacuole displacing nuclei peripherally (H&E, 40x).

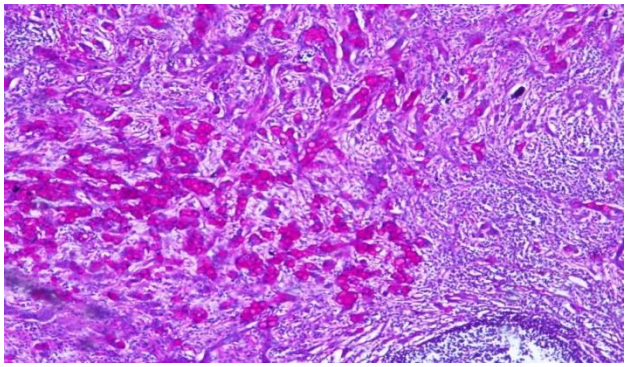

Figure 4: PAS stain show intracytoplasmic mucin in signet ring cells in magenta colour (PAS, 10x).

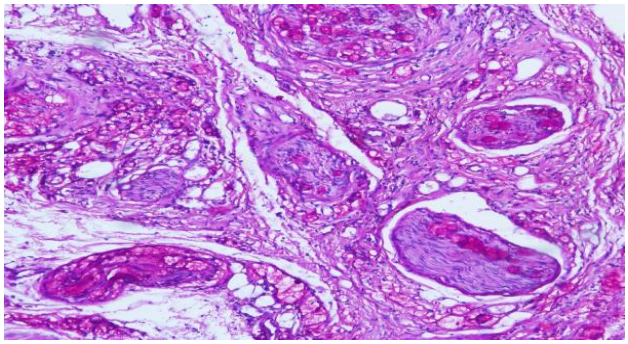

Figure 5: PAS stain show signet ring cells nerves and vessels in magenta colour (PAS, 10x).

DISCUSSION

Only few cases of appendiceal signet ring cell carcinomas have been described in the literature. Primary adenocarcinoma of the appendix is a rare malignancy of the gastrointestinal tract.[1] The demographic characteristics of patients with cancer of the appendix vary by histology. According to report, those diagnosed with malignant carcinoids are significantly younger (mean age, 38 years) than those diagnosed with any of the other cancer types.[1] The mean age of patients at diagnosis of mucinous adenocarcinoma, colonic type

adenocarcinoma, and signet ring cell carcinoma is approximately 60 years, 62 years, and 58 years, respectively, and an equal number of males and females developed goblet cell carcinoid, mucinous adenocarcinoma, and signet ring cell carcinoma, except for colonic adenocarcinoma which had a male predominance.[1] Our patient was 55 year male which is near above data.

According to WHO book, only appendiceal adenocarcinoma containing more than 50% of SRCC elements is called appendiceal SRCC.[3] Signet ring cell carcinomas, usually frequent in stomach and intestine, are adenocarcinomas with mucus-producing tumour cells. Our case was composed of SRCC element (60%); thus our case fulfils the criteria of primary appendiceal SRCC. The primary SRCC of the current case involved two third of appendix with metastasis to the lymph node. The present SRCC of the appendix seems to be the primary site since no tumours other than the appendiceal tumour were found in the body by USG. The present primary SRCC of appendix clinically manifested as acute appendicitis. The lumen of appendix was obliterated; Furthermore, the depth of invasion of the current SRCC was upto serosa, and neuro-lymphovascular permeation was seen.

Most appendiceal cancers are low-grade neoplasms that are typically relatively indolent. The overall 5-year survival rate for mucinous appendiceal adenocarcinomas was 20.5%.[4] According to report of McCusker, except for signet ring cell carcinoma and malignant carcinoid, the histologic type does not have a significant impact on survival.[1] In addition, the extent of disease at diagnosis is a more important predictor of survival than histology. In a study by McGory et al, poorly differentiated adenocarcinoma and signet ring cell carcinoma of the appendix had the highest proportion of distant disease with a 5-year survival rate of 7%.[5] Therefore, signet ring cell carcinoma may be a separate tumour type in the appendix that should be considered apart from other carcinomas, largely because of its poor prognosis. Right hemicolectomy is considered the optimal treatment for most histologic types of primary appendiceal carcinoma even in the presence of perforation and in Dukes A tumors.[6] While some surgeons suggest that a simple appendectomy is sufficient for tumours exhibiting only local disease, many studies have shown that there is a clear survival benefit to the addition of a hemicolectomy.[6,7] The treatment options for metastatic disease include systemic chemotherapy alone, hyperthermic intraoperative intraperitoneal chemotherapy, cytoreductive surgery with peritonectomy, and combination of treatments. Of those treatment options, cytoreductive surgery and hyperthermic intraperitoneal chemotherapy have recently become the treatment of choice for metastatic diseases at most large centers.[8] Our patient was managed by post appendectomy hemicolectomy. After passing 5 month of hemicolectomy patient have not any clinical problem related to cancer and recurrence.

CONCLUSION

Signet-ring cell carcinoma (SRCC) of vermiform appendix is extremely rare which clinically presenting usually as acute appendicitis. It had the highest proportion of distant disease and poor prognosis. However it is diagnostic challenge to diagnose preoperatively but has great importance in operative decision.

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