

A Rare Case of Primary Peritoneal Psammocarcinoma Presenting as Acute Intestinal Obstruction and Ascites

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ABSTRACT

We report a rare case of primary peritoneal psammocarcinoma (PPP) in a 60-year-old female patient who presented to surgical emergency with complaints of inability to pass feces and flatus for last 3 days along with pain and distension of abdomen. On clinical examination there were features of acute intestinal obstruction. Contrast CT of abdomen was suggestive of intestinal obstruction along with a stricture at the recto-sigmoid junction. On exploration there was a growth compressing the recto-sigmoid junction from the serosal surface. Anterior resection of the recto-sigmoid was done with negative gross margins. Histopathological examination of the resected specimen led to a diagnosis of primary peritoneal psammocarcinoma because of a solitary peritoneal lesion having abundant psammoma body formation and normal ovaries visualized on radiology and per-operatively. The diagnosis was further confirmed by histochemical tests and immunohistochemistry along with normal CA-125 levels in the patient.

Keywords: Peritoneal Psammocarcinoma, Intestinal obstruction, Ascites

INTRODUCTION

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PPrimary peritoneal psammocarcinoma (PPP) is a very rare malignant tumor of the peritoneum characterized by extensive psammoma body formation.^[1] Its behaviour is indolent and similar to the low malignant potential tumors of the ovary.^[2] Per-operatively Primary peritoneal psammocarcinoma is differentiated from primary ovarian carcinoma if there is presence of peritoneal lesions in the absence of gross involvement or only superficial involvement of the ovaries.^[3] These tumors may at times behave aggressively with recurrence or metastases but many cases are incidentally discovered during laparotomy done for other conditions.^[4,5] Hence, though rare but this entity should also be considered in the differential diagnoses of peritoneal carcinomatosis.

CASE REPORT

A 60-year-old female patient presented to surgical emergency with complaints of inability to pass feces and flatus for last 3

days associated with pain and distension of abdomen. She also gave history of similar episodes in the past one year, which were relieved on conservative management, but she was never investigated for the same. On presentation, she was hemodynamically stable except for mild tachycardia. Per-abdominal examination revealed abdominal distension with exaggerated bowel sounds. Radiographs of the abdomen showed dilated bowel loops with air fluid levels. Double contrast CT abdomen was suggestive of an annular growth at the recto-sigmoid junction (Figure 1).

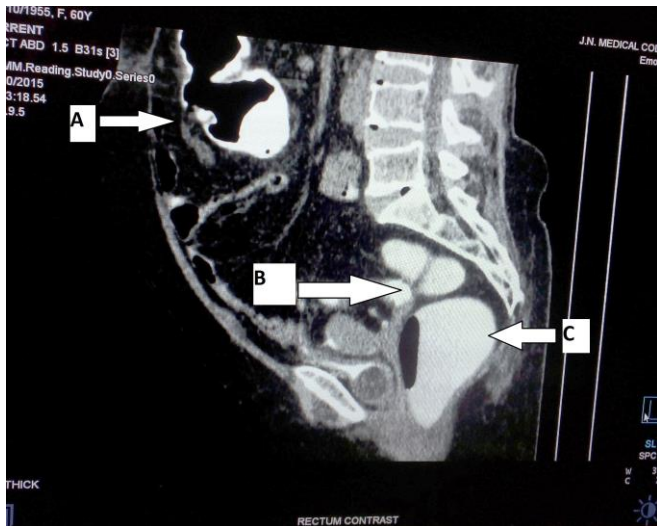


Figure 1: Sagittal section of the contrast CT abdomen showing (a) Cut-off of the contrast material in the region of sigmoid colon suggesting intestinal obstruction (b) Filling defect of contrast material and malignant growth at recto-sigmoid junction (c) Rectal contrast with abrupt tapering.

Patient was planned for exploratory laparotomy and on exploration, there was a growth compressing the recto-sigmoid junction from the serosal surface along with free fluid in the pelvis. Ascitic fluid was sent for cytological analysis. Anterior resection of the recto-sigmoid was done with negative gross margins and specimen was sent for histopathological examination (Figure 2a&b).

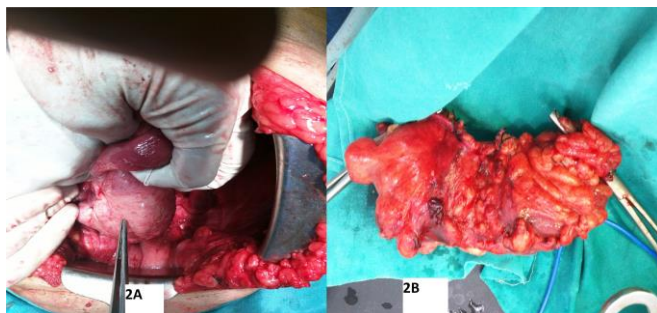


Figure 2a: Per-operative Photograph of the patient showing malignant growth at the recto-sigmoid junction with nodules over the serosal surface.
Figure 2b: Photograph of the resected specimen with nodules over the serosa and malignant growth

Ascitic fluid cytology showed 3-D clusters and sheets of malignant cells with eosinophilic cytoplasm, raised nucleo-cytoplasmic ratio, pleomorphic nuclei with clumped chromatin and prominent nucleoli in some cells (Figure. 3a). Gross specimen showed a stricture in sigmoid colon with a whitish growth 2x2 cm on the serosal surface. Hematoxylin and Eosin stained sections from the area of growth showed normal intestinal mucosal glands, submucosa and muscular layer but serosal surface showed malignant glands lined by moderately pleomorphic round to ovoid cells with raised nucleo-cytoplasmic ratio and hyperchromatic nuclei. Abundant psammoma bodies were also seen (Figure. 3b&c). Owing to the absence of any other primary lesion and normal ovaries a diagnosis of primary psammomatous adenocarcinoma of the peritoneum was made. The 10 lymph nodes dissected out from the mesocolic fat showed no malignant deposits. Serum CA-125 of the patient was 23U/ml which was within the normal limits. Histochemistry with Periodic acid Schiff (PAS) stain showed diastase resistant PAS positivity. Further immunohistochemistry with CK7 was done which came out to be positive (Figure 3d) CK 5/6 was negative, hence confirming the diagnosis of primary psammomatous adenocarcinoma of the peritoneum stage IIIB (TNM/FIGO).

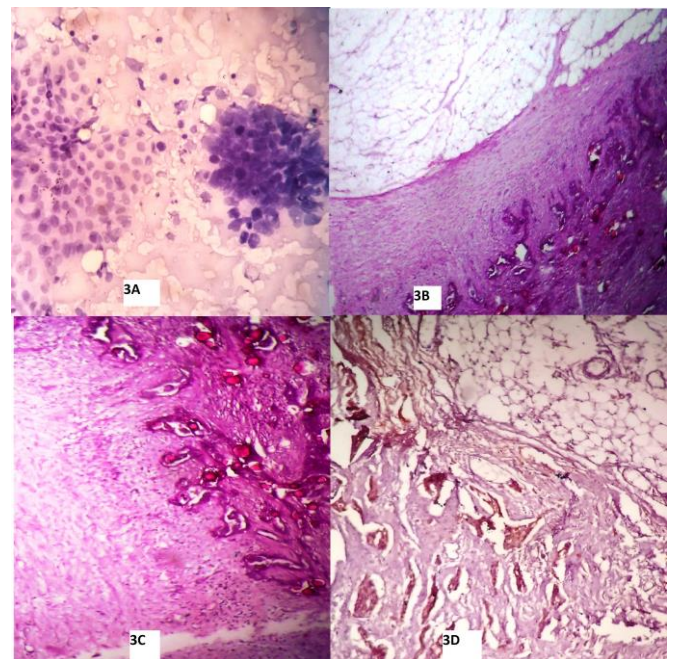


Figure 3a: Hematoxylin & Eosin X 400: cytological smear from Ascitic fluid showing sheets and clusters of malignant cells favouring adenocarcinoma. Some singly dispersed malignant cells also seen on a proteinaceous background.
Figure 3b: Hematoxylin & Eosin X 100: section showing small malignant glands in the serosal layer with abundant psammoma body formation.
Figure 3c: Hematoxylin & Eosin X 400: high power view of the glands lined by moderately pleomorphic cells along with psammoma bodies.
Figure 3d: CK7 X 400: The glandular cells show cytoplasmic positivity CK7.

DISCUSSION

Primary psammocarcinoma is a rare epithelial tumour which arises from the ovary or peritoneum. It was first described by Kettle in 1916 and later on Gilks pathologically classified this entity in 1990.^[6] Clinically they are similar to borderline serous tumours with a less aggressive and comparatively favorable outcome.^[7] As described by Gilks et al, the morphological characteristics for the diagnosis of psammocarcinoma include invasive lesions of the ovarian stroma, blood vessels, intraperitoneal viscera or peritoneum with small nests of epithelial cells not more than 15 cells in diameter having moderate nuclear atypia and abundant psammoma body formation in at least 75% of the papillae or epithelial cell nests.^[8] These criteria were further revised by Chen et al emphasizing the presence of involvement of intra-abdominal viscera or presence of an invasive growth in the peritoneum.^[9] Based on these criteria our case was diagnosed to be primary peritoneal psammocarcinoma. The abundant psammoma bodies (50-100 microns in diameter) seen in such cases are a result of dystrophic calcification leading to formation of calcified spherules with concentric laminations formed by deposition of hydroxyapatite in degenerating cells.^[10] They may also be seen in papillary thyroid cancer, meningioma, ovarian tumours or some gastrointestinal tumours like duodenal carcinoids or gastric adenocarcinoma.^[11, 12]

Low-grade ovarian tumours, including peritoneal psammocarcinoma may be attributed to mutations in BRAF gene leading to defect in kinases regulated by RAS. Similar mutations may also be seen in certain thyroid or colorectal malignancies.^[6, 13]

Patients are usually older with a mean age of incidence of around 55 years.^[8] Our patient was also an old lady of 60 years who presented with complaints of inability to pass feces and flatus for last 3 days and pain and swelling over abdomen. Moreover, she also complained of vague abdominal discomfort for past 1 year. Most of the patients with this entity present like FIGO stage 3 ovarian borderline or malignant tumours and usually have similar complaints of abdominal discomfort or swelling.^[11] Some patients may have moderate elevations of CA-125, high levels of this marker signify an aggressive behavior of the tumor.^[14] In our patient CA-125 was not raised.

Peritoneal psammocarcinoma needs to be differentiated from the aggressive peritoneal serous adenocarcinoma which have solid growth, high grade cell atypia and inconspicuous psammoma body formation.^[6] Another rare differential diagnosis is calcified leiomyomatosis peritonealis disseminata which affects females of reproductive age group who have undergone laparoscopic myomectomy.^[14] However it is ruled out in our case as the patient is postmenopausal without any history of laparoscopic uterine surgery. Benign lesions like cystadenofibromas and endosalpingiosis may also pose diagnostic difficulties but they usually have no or minimal

cytologic atypia and inconspicuous psammoma body formation.^[8]

Once diagnosed, patients of primary peritoneal psammocarcinoma need optimal surgical debulking in the form of hysterectomy with bilateral salpingo-oophorectomy and omentectomy. Beneficial role of adjuvant chemotherapy in PPP is still not proven but it could be given. Owing to the expression of hormone receptors in these tumours, tamoxifen may be used as an alternative and effective approach in some cases.^[6, 8]

CONCLUSION

Primary peritoneal psammocarcinoma is a rare malignancy of the peritoneum with excessive psammoma body formation which usually has an indolent course like borderline ovarian serous tumours but may behave aggressively at times. Patients are treated as FIGO stage III ovarian tumours with optimal debulking. Role of adjuvant chemotherapy is controversial however; hormone therapy with tamoxifen may have a favorable role. Patients should be religiously followed –up to look for any recurrence. In our case patient was followed-up and she was doing well after 6 months of surgery.

What this study adds

1. What is known about this subject?

Primary peritoneal psammocarcinoma is a very rare malignant tumor of the peritoneum in which there is extensive psammoma body formation. Its behavior is similar to low malignant potential tumors of the ovary and it is also considered one of the differential diagnoses of peritoneal carcinomatosis.

2. What new information is offered in this study?

The patient presented with features of intestinal obstruction and was suspected to be having colorectal carcinoma on the basis of CT scan but actual pathology diagnosed after histopathological examination was Primary peritoneal psammocarcinoma. This type of presentation of Primary peritoneal psammocarcinoma is very uncommon.

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