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A Rare Case of Pseudomyxoma Peritonei: Where did its Culprit Originate?

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Abstract

Pseudomyxoma peritonei refers to accumulation of mucin in the peritoneal cavity leading to a gelatinous form of ascites. With its incidence ranging between 1-4 out of a million per year, it is an extremely rare entity to be encountered by surgeons and gynaecologists. We report a case of a 55-year-old patient who presented with complaints of abdominal distension and dull abdominal pain for 2 months. On examination, abdomen appeared grossly distended, a soft cystic mass was felt arising from pelvis and extending up to the epigastrium. Patient underwent an exploratory laparotomy followed by total abdominal hysterectomy with left salpingo ovariotomy and right salpingoopherectomy with infracolic omentectomy. Intra operatively, it was observed that entire intra-peritoneal cavity was filled with mucin, a large multiloculated cyst engulfing the left ovary was noted, on further examination of the bowel a thickened appendix with mucinous deposit over it was identified for which appendectomy was performed. On histopathology, pseudomyxoma peritonei was confirmed with mucinous carcinoma of appendix and mucinous cystadenocarcinoma of left ovary. Furthermore, on immunohistochemistry testing it was revealed two independent primary malignancies of left ovary and appendix were coexisting.

Keywords: Appendiceal carcinoma; Mucinous cystadenocarcinoma, Appendectomy, Ascites

Introduction

The incidence of pseudomyxoma peritonei is minuscule, ranging from 1-4 out of a million per year [1]. It is recognized by diffusely spread intraabdominal gelatinous ascites along with mucinous deposits implanted over peritoneal surfaces. It's most frequent originating site is known to be appediceal neoplasm but other recognized sites include stomach, small and large intestine, fallopian tubes and ovaries. On the other hand, large mucinous ovarian neoplasms can be primary tumors or metastatic carcinomas from gastrointestinal tract [2]. To differentiate and determine origin is essential for further course of management and prognostication. Clinical presentation spectrum of pseudomyxoma peritonei ranges from asymptomatic incidental finding in early-stage disease to symptomology constituting of abdominal distension, pain, indigestion, presence of a pelvic mass or presence of obstructive bowel symptoms in advanced disease.

Case Report

A 55-year-old female patient presented with complaints of abdominal distention and dull abdominal pain for 2 months. On examination, abdomen appeared grossly distended, a soft cystic mass was felt arising from pelvis

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and extending up to the epigastrium. There was no associated lymphadenopathy noted. Her biochemical tests revealed raised Ca-125 and CEA levels of 114.3 U/ml and 16.4 ng/ml respectively. Furthermore, on imaging, Computed Tomography scan showed a large multiloculated cystic lesion arising from left ovary with evidence of gross ascites (Figure 1).

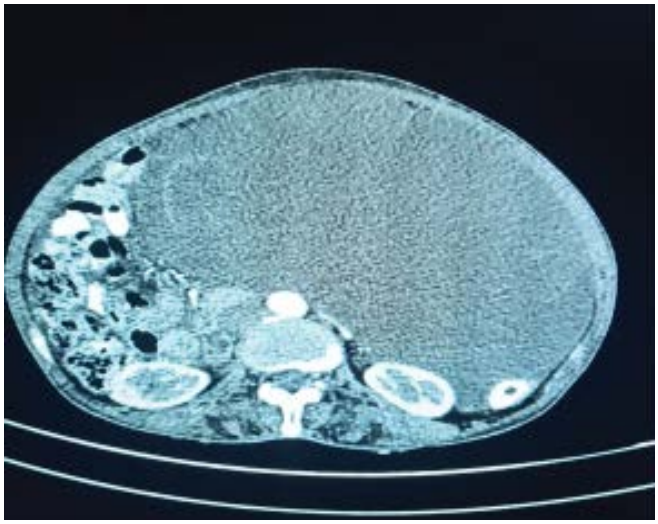

Figure 1: CT scan of the abdomen showing gross presence of large left ovarian multilocular cyst with gross ascites.

We decided to perform an exploratory laparotomy upon which intraoperatively, it was observed that entire intra-peritoneal cavity was filled with mucin, a large 15 × 15 × 10 cm multiloculated cyst engulfing the left ovary was noted, on further examination of the bowel a thickened appendix with mucinous deposit over it was identified (Figure 2). We went ahead and performed a total abdominal hysterectomy with left salpingo ovariotomy and right salpingoopherectomy with infra colic omentectomy whereas an appendectomy was performed by a general surgeon in the same sitting.

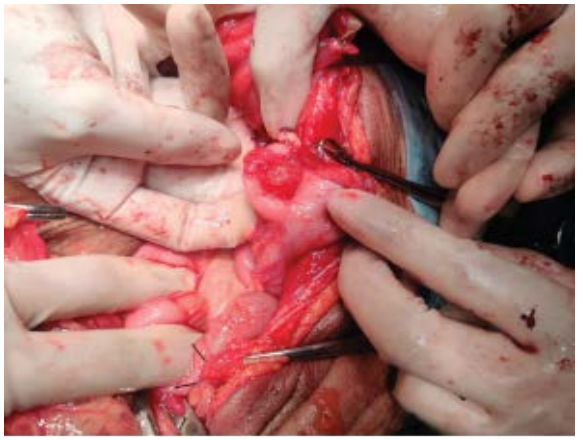

Figure 2: Intraoperatively seen enlarged appendix with mucinous deposits.

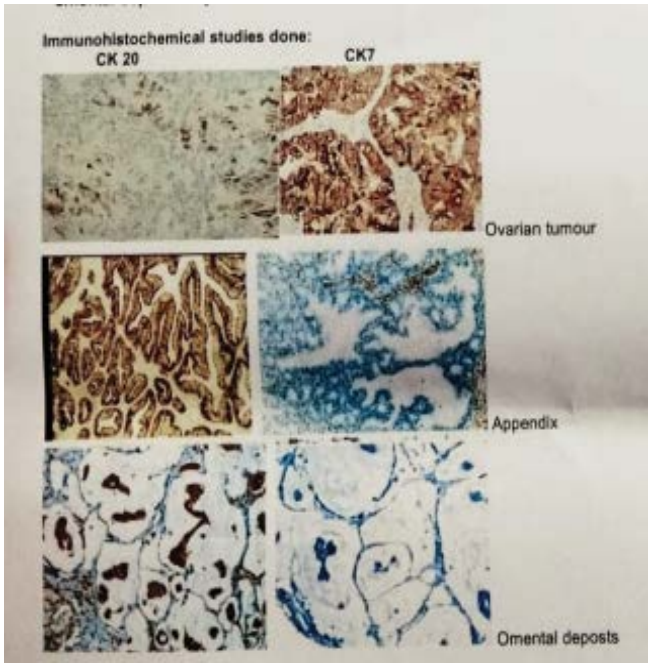

Figure 3: Immunohistochemistry results showing ovarian tumour strongly positive for CK 7 and focally positive for CK 20; Appendiceal tumour and omental deposits showing strong positivity for CK 20 and negative for C7.

On histopathology, impression of pseudomyxoma peritonei was confirmed. There was also mucinous carcinoma of appendix with mucinous cystadenocarcinoma of left ovary with mucinous carcinomatous deposits on both right and left fallopian tubes.

On IHC testing it was revealed that two independent primary malignancies of left ovary and appendix were coexisting. The ovarian tumor was strongly positive for CK7 and focally positive for CK 20 whereas appendiceal and omental deposits were positive for CK20 and negative for CK7 (Figure 3). Post operative period of this patient in our hospital was uneventful, she was discharged on 7th post operative day and further referred to a gynaecologic oncologist to discuss prospective treatment course.

Conclusion

Appendiceal mucinous neoplasms (AMNs) are unusual tumours accounting for less than 1% of all cancers [3] whereas mucinous ovarian cystadenocarcinomas constitute 12% of ovarian malignancy [4]. The occurrence of these tumours synchronously along with the presence of pseudomyxoma peritonei is worth reporting because of its pure rarity. In literature most cases show a primary appendiceal malignancy which has metastasized to the ovaries. It is imperative to stress upon the role of immuno histochemistry in identifying the distinct origin of the two tumours. We further lay emphasis on the need for appendicectomy even in cases where appendix looks grossly normal as microscopic involvement

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may be present. Given the simplicity of the procedure with no additional morbidity it must be done in cases with mucinous tumours of the ovary to improve prognosis.

Compliance with ethical standards

Conflict of interests: The authors declare they have no conflict of interest.

Informed consent: Informed consent was taken from the participant who was included in the study.

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