

Primary Peritoneal Papillary Serous Cystadenocarcinoma - A Rare Malignancy

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Rezumat

Chistadenocarcinom papilar seros peritoneal primar - o formă rară de neoplazie

Carcinomul papilar seros peritoneal este o formă rară de neoplazie primară, ce afectează difuz peritoneul abdominal și pelvin. Cancerele ovariene epiteliale și cancerele peritoneale primare au originea în epiteliul germinal comun, dezvoltat din epiteliul celomic. Datorită unei origini embrionare comune a ovarului și peritoneului, cancerele de ovar și carcinomul peritoneal primar prezintă multiple similarități histologice. Cu toate acestea, incidența carcinomului seros peritoneal primar este mult mai scăzută decât a cancerului ovarian epitelial și se întâlnește cel mai frecvent la femei în vârstă. Stadiile incipiente de boală pot fi asimptomatice; simptomele apărute în stadiile avansate includ: distensie abdominală, formațiune tumorală abdominală, durere abdominală cu caracter nespecific, vărsături și dispnee, ca urmare a ascitei masive. Pacienții diagnosticați cu carcinom papilar seros peritoneal primar sunt tratați pe baza aceleiași stadializări și aceluiași abordări chirurgicale și chimioterapice aplicate în cazul cancerului ovarian epitelial datorită similitudinilor manifestărilor biologice.

Cuvinte cheie: carcinom papilar seros peritoneal primar, cancer ovarian epitelial, epiteliu celomic

Abstract

Primary peritoneal papillary serous carcinoma is a rare primary malignancy diffusively involving the peritoneum of abdomen and pelvis. Epithelial ovarian cancers and primary peritoneal cancers arise from the common germinal epithelium which develops from the coelomic epithelium. Due to a common embryonic origin of the ovary and the peritoneum, carcinoma of the ovary and primary peritoneal carcinoma have much histological similarity. However the incidence of Primary peritoneal serous carcinoma is considerably lower than that of epithelial ovarian cancer and is mostly seen in elderly women. Early stages of this disease may be asymptomatic; symptoms of the advance stages of the disease include abdominal distention, abdominal lump, non-specific abdominal pain, vomiting and dyspnoea all as a result of massive ascites. Patients diagnosed with primary peritoneal papillary serous carcinoma are treated using the same staging, surgical and chemotherapeutic approach as epithelial ovarian cancer because of the similarities in biological behavior.

Key words: Primary peritoneal papillary serous carcinoma, epithelial ovarian cancer, coelomic epithelium,

Introduction

Primary peritoneal papillary serous carcinoma is a rare primary malignancy diffusively involving the peritoneum of abdomen and pelvis and was first described in 1959 by Swerdlow as a mesothelioma of the pelvic peritoneum. (1)

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Case report

A 43 year female presented to the surgical clinic with complain of gradually progressive painless lump in the right lumbar region without any associated symptoms. General examination of the patient was unremarkable. On abdominal examination an ill defined mass was present in the right lumbar region of size 10 x 10 cm, non tender and firm in consistency with smooth surface and not moving with respiration. Routine hematological investigation, renal function tests and liver function test did not showed any abnormality.

Oral and intravenous contrast enhanced CT scan of abdomen was done which revealed a well defined hypodense lesion with peripheral wall calcification and thin internal septations of size 10 x 8.2 x 8.1 cm present over the right kidney, pushing the kidney postero-laterally and head of pancreas towards then left side. Fat plane with liver was well maintained. (Fig. 1)

Exploratory laparotomy was done. Intraoperative a cyst of size 10x10 cm was found in the right lumbar region loosely adhered to right kidney, duodenum and right lobe of liver. Ovaries were found to be normal. Complete excision of the intact cyst was done. (Fig. 2) Patient had an uneventful post operative recovery.

Histopathology of the specimen showed a moderately differentiated primary peritoneal papillary cystadenocarcinoma. (Fig. 3) Serum CA125 level was planned and was found to be in values of 10.08 U/ml (Normal 0-35U/ml). Patient was followed up for Chemotherapy.

Discussion

Primary peritoneal papillary serous carcinoma is a rare tumor which spreads widely on the peritoneal surfaces involving mostly the omentum with minimal or no ovarian involve-

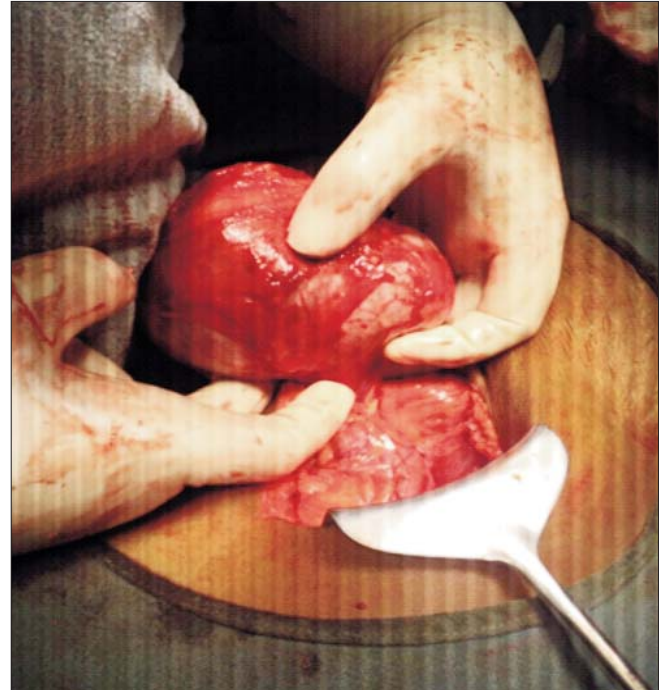


Figure 2. Cyst of size 10x10 cm was found in the right lumbar region loosely adhered to right kidney, duodenum and right lobe of liver

ment. Epithelial ovarian cancers and primary peritoneal cancers arise from the common germinal epithelium which develops from the coelomic epithelium. Due to a common embryonic origin of the ovary and the peritoneum, carcinoma of the ovary and primary peritoneal carcinoma have much histological similarity. Other histologic variants of the non serous primary peritoneal carcinomas have also been reported; specifically, endometrioid, clear cell, mucinous, Brenner tumor, and mixed müllerian tumors; but non serous and serous

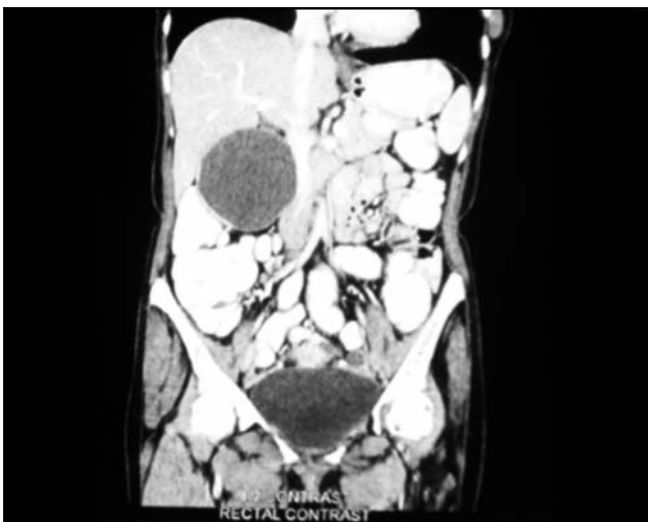


Figure 1. CECT abdomen was done which revealed a well defined hypodense of size 10 x 8.2 x 8.1 cm present between right kidney, pancreas & right lobe of liver

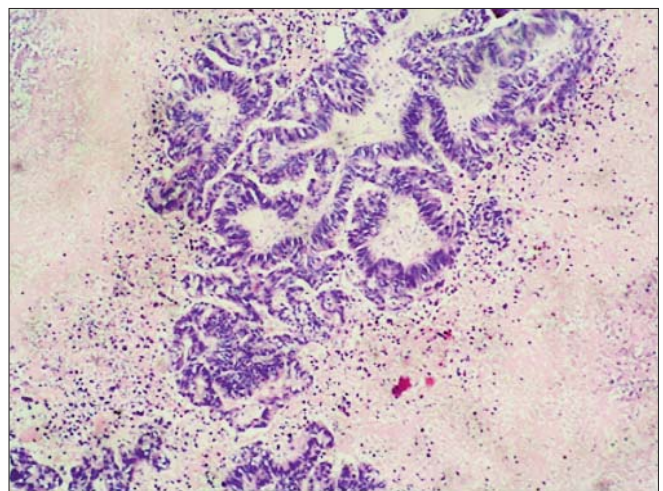


Figure 3. Histopathology of the specimen showed a moderately differentiated primary peritoneal papillary cystadenocarcinoma

tumors appear to be similar with regard to prognosis and response to therapy (2) (3) However the incidence of Primary peritoneal serous carcinoma is considerably lower than that of epithelial ovarian cancer, 6.78 cases per million versus 120.5 cases per million, respectively. (4) This entity has also been reported under various names including extraovarian primary peritoneal carcinoma, extraovarian müllerian adenocarcinoma, multiple focal extraovarian serous carcinoma, normal-sized ovary carcinoma syndrome, primary peritoneal serous carcinoma. (5, 6)

Primary peritoneal papillary serous carcinoma is mostly seen in elderly women. However, rare cases have been reported in children and in males also. (6, 7, 8) Early stages of this disease may be asymptomatic; symptoms of the advance stages of the disease include abdominal distention, abdominal lump, non-specific abdominal pain, vomiting and dyspnoea all as a result of massive ascites. Imaging of abdomen by sonography, contrast enhanced CT scan and MRI abdomen will help in diagnosing this condition showing presence of ascites, abdominal masses “omental cakes” and tumour nodules on various peritoneal surfaces with normal size ovaries. Fine needle aspiration cytology may sometimes help in making the diagnosis. The diagnostic criteria of this condition to differentiate it from primary serous carcinoma of ovary has been defined by the Gynecology Oncology Group includes (1) ovaries must be normal size or enlarged as result of benign process (2) extraovarian involvement must be greater than the surface involvement of either ovary (3) ovarian involvement must be absent, confined to the ovarian surface epithelium without stromal invasion, or involve the cortical stroma with a maximal tumour dimension of less than 5x5 mm. (2) However both the tumours have similar elevation in levels of tumour makers CEA and CA-125. (9)

Patients diagnosed with primary peritoneal papillary serous carcinoma are treated using the same staging, surgical and chemotherapeutic approach as epithelial ovarian cancer because of the similarities in biological behavior. The management of consists of combining optimal surgical debulking followed by similar chemotherapy regimen as used for epithelial ovarian cancer. Post treatment patients can

evaluated for improvement of symptoms clinically, radiologically, serum CA-125 level. Prognosis and survival data is still limited. Survival rates however may be less than survival rates for similar stage ovarian cancer (10, 11)

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