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Pathology

Primary Peritoneal Carcinoma – A Rare Challenging Cytological Diagnosis

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Abstract **Case Report**

Primary peritoneal carcinoma (PPC) is a rare multicentric malignant tumour of the peritoneal cavity often morphologically indistinguishable from primary ovarian serous carcinoma and carcinoma of the fimbrial end of the fallopian tube. A correct differential diagnosis of PPC is highly challenging. The current case report describes the case of PPC in a 70 year old female presented with abdominal distension and elevated CA 125 levels. The case report highlights the importance of high index of suspicion, role of cytology and imaging techniques in the diagnosis of this rare entity.

Keywords: Primary peritoneal carcinoma, primary ovarian serous carcinoma, Cytological diagnosis.

INTRODUCTION

Primary peritoneal carcinoma (PPC) is a rare multicentric malignant tumour of the peritoneal cavity often morphologically indistinguishable from primary ovarian serous carcinoma and carcinoma of the fimbrial end of the fallopian tube. The cell of origin of PPC is the peritoneal embryonic nests of mullerian cells [1]. It accounts for 10% of pelvic serous carcinoma and it is almost exclusively found in women. The tumor appears during adulthood with a mean age of 60 years.

PPC greatly involves the peritoneal surface extensively and lacks the ovarian surface involvement proven either by imaging techniques or microscopic examination [2]. The current case report describes the case of PPC in a 70 year old female presented with abdominal distension and elevated CA 125 levels. Imaging findings revealed peritoneal nodules with no identifiable primary site. The case report highlights the importance of high index of suspicion, role of cytology and imaging techniques in the diagnosis of this rare entity. To our best knowledge, only very few cases of primary peritoneal carcinoma has been described in the literature.

CASE HISTORY

A 70 year old female presented with history of early satiety and abdominal distension with loss of weight and appetite for 1month. CT Abdomen revealed gross ascitis with omental, peritoneal thickening and nodules. In addition, there was a hiatus hernia with circumferential wall thickening of esophagus and focal lesion in the liver measuring 1.8 x 1cm suggestive of metastasis. Upper gastrointestinal endoscopy was normal. Serum marker CA 125 was more than 600 (normal range, 0–35 U/ml).

Ascitic fluid tapping was done for further evaluation which yielded 120 ml of pale yellow turbid fluid. The sample was centrifuged for cytodiagnosis and cell block.

The cytological smears were highly cellular and showed papillaroid, three dimensional and loosely cohesive clusters, acinar, sheets and individual polygonal cells with enlarged nuclei, increased nucleocytoplasmic ratio, moderate nuclear anisokaryosis and moderate cytoplasm. Few cells showed cytoplasmic vacuolations. Background showed RBCs and scattered inflammatory cells and few reactive mesothelial cells. Histopathogical sections from the cell block of the aspirated fluid showed the similar morphology. Based on the microscopic findings, clinical details and imaging findings, a possibility of a primary peritoneal carcinoma was given as diagnosis. Further the patient was referred to higher centre where IHC confirmation was done on cell block.

Legends for Figures

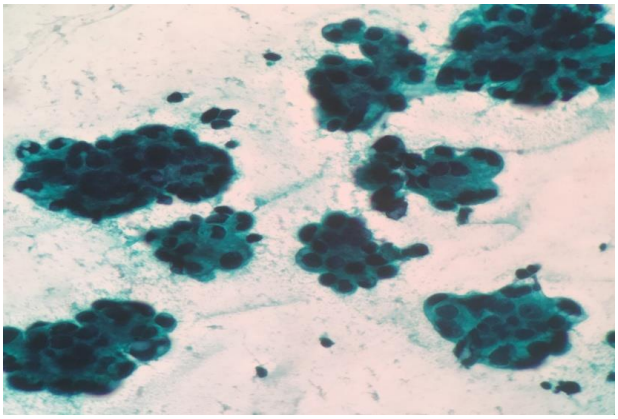

Figure 1: Cytocentrifuged smears revealing high cellularity, with clusters and papillary fragments of highly pleomorphic malignant cells (Pap, 400 x)

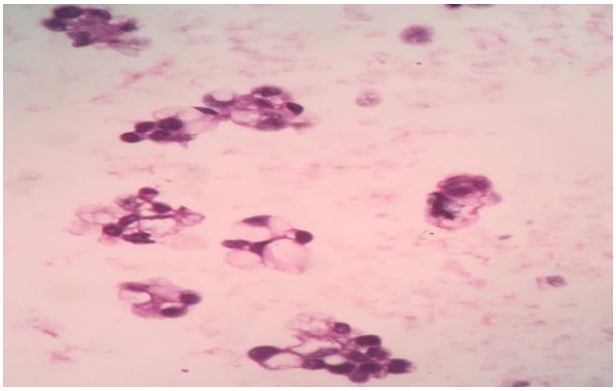

Figure 2: Cell block section showing glandular pattern of malignant cells with cytoplasmic vacuolations.(Hematoxylin and Eosin, x 400x)

DISCUSSION

Peritoneal neoplasms can be primary or secondary based on the site of origin of the tumor. Primary peritoneal malignancies are rare entities which includes malignant mesothelioma, serous carcinoma and sarcoma. The entity of primary peritoneal carcinoma was first described by Swerdlow in 1959 as mesothelioma of the pelvic peritoneum. The secondary mullerian nature of female peritoneum including the pelvic mesothelium was further explored by Lauchlan in 1972. Due to invagination of the coelomic epithelium, the epithelial lining of ovary and pelvic peritoneum share a common embryonic origin and may undergo independent malignant transformation [1].

The median age of presentation of PPC is 64 years and usually higher than that of patients with epithelial ovarian cancer [3]. The patients usually presents with abdominal fullness, dull aching abdominal pain and abdominal mass due to massive ascites [4]. A preoperative diagnosis of primary peritoneal carcinoma poses a great diagnostic challenge and is often misdiagnosed as peritoneal carcinomatosis from a gastrointestinal or ovarian malignancy [5].

Imaging techniques like PET/CT which define the extent of metabolically active disease and distant metastasis are highly beneficial in differentiating PPC from epithelial ovarian cancer [6].

Few studies have highlighted the utility of endometrial cytological sampling in detecting ovarian, tubal and peritoneal high grade serous carcinomas in patients who present without any detectable pelvic mass [7]. Hattori *et al.*, described 8 cases of PPC sporadically developed in the peritoneal or extra peritoneal cavity with or without malignant ascites. The criteria developed Gynecologic Oncology Group (GOG) for the diagnosis of PPC includes a normal sized ovary, greater involvement of the extra ovarian sites compared to the surface of either ovary, uninvolvement of the ovary microscopically and also the serous nature of the lesion confirmed cytologically and histopathologically [8]. Ayhan *et al.*, [9] revealed similar clinical characteristics between PPC and epithelial ovarian cancer, Huang *et al.*, found [10]. Allelic loss on chromosome 6q which can contribute to the multifocal nature of PPC.

Sorensen *et al.*, [11] considered PPC and epithelial ovarian cancer as separate entities with different carcinogenic pathways as they observed differences in the risk factor profile, prognostic and molecular patterns of these two entities. Surgical resection, platinum based chemotherapy; intraoperative chemotherapy provides the optimal treatment modalities for the patients. However, published survival data from the few reported cases is 11-17 months [12].

CONCLUSION

This condition is clinically and histologically similar to advanced stage serous ovarian carcinoma extra ovarian primary peritoneal carcinoma is a rare malignant epithelial tumor, which arises from the peritoneal lining with minimal or no ovarian involvement. This condition is a diagnosis of exclusion with the absence of other identifiable primary site. It is to be differentiated from peritoneal carcinomatosis and malignant mesothelioma with adequate clinical detail and immunohistochemistry.

SOURCES OF SUPPORT: NIL
CONFLICTS OF INTEREST: NIL

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