Primary papillary serous carcinoma of the peritoneum

INTRODUCTION

Primary papillary serous carcinoma of the Peritoneum (PSCP) is a primary tumor of the peritoneum that diffusely involves the peritoneal surface but spares the ovaries.\(^{[1,5,4]}\) The closest differential for this entity is serous ovarian papillary carcinoma.\(^{[1]}\) These cannot be distinguished on the basis of histopathology and immunotyping. The only imaging distinction between these two conditions is non-involvement of the ovaries in PSCP.

We present a case of PSCP in a 72-year-old female which was clinically and histologically indistinguishable from papillary serous carcinoma of the ovary, but radiologically it can be differentiated on the basis of non-involvement of the ovaries.

CASE REPORT

A 72-year-old female patient presented to our outpatient department with complaints of gradual-onset lower abdominal discomfort with distension for the past 1 year. No previous history of post-menopausal bleeding, fever or pelvic discharge was present. No family history of primary pelvic malignancy was revealed. On bimanual pelvic examination, nodules and irregularity were palpated in the rectouterine pouch; however, bilateral adnexa, uterus and cervix appeared unremarkable on clinical examination. Serum CA-125 was found to be elevated.

The patient was referred to our department for further imaging evaluation. Grey scale ultrasound revealed gross ascites and few well-defined hypoechoic nodules along the mesentery and in the rectouterine pouch. The uterus appeared atrophied and both adnexa appeared unremarkable.

Further, the patient underwent cross-sectional imaging. The non enhanced computed tomography (NECT) scan showed few ill-defined confluent lesions in the omentum with no evidence of internal calcification. No evidence of any bony sclerotic or lytic lesion is noted [Figure 1a].

Contrast-enhanced computed tomography demonstrated a heterogeneously enhancing ill-defined confluent mass-like thickening in the omentum s/o omental caking [Figure 1b]. Multiple areas of focal enhancing thickening and nodules were noted along the mesentery and peritoneum predominantly in the rectovaginal pouch and urovesical pouch, with few of them in close relation with the anterior and posterior walls of the atrophied uterus. Bilateral adnexae showed no evidence of a focal lesion with no radiological signs of ovarian enlargement or ovarian capsular breech or invasion. No evidence of significant lymphadenopathy was noted in the abdomen [Figure 2a and b].

For better evaluation of the ovaries, non-contrast magnetic resonance imaging of the pelvis was
performed. Axial fat spin echo T2-weighted images confirmed normal appearing ovaries bilaterally with absence of gross pathology [Figure 3a and b].

Cytopathological examination of the sample obtained from the thickened pelvic peritoneum and omental mass (Papanicolaou stain and hematoxylin and eosin [H and E] stain) showed clusters of malignant cells having eccentric round vesicular nuclei and prominent nucleoli [Figure 4a]. In addition, cells showed papillaroid, adenoid and solid sheet-like arrangement [Figure 4b] with multinucleated giant cells [Figure 4c].

DISCUSSION

Primary papillary serous carcinoma of the peritoneum (PSCP) is defined as a primary tumor of the peritoneum exclusively found in women that diffusely involves the peritoneal surface but spares the ovaries. They are believed to arise from the embryonic nests of the Müllerian cells present in the peritoneum and are derived from the embryonic coelomic epithelium. The closest differential for this entity is serous ovarian papillary carcinoma. These cannot be distinguished on the basis of histopathology and immunotyping. The only imaging distinction between these two conditions is non-involvement of the ovaries in PSCP. However, it is now increasingly believed that the distinction between the two conditions may not be necessary as both of them have similar management approaches. More importantly, PSCP must be differentiated from more sinister pathologies like peritoneal carcinomatosis due to other primaries, lymphomatosis and mesothelioma because all of them have different management approaches.

To distinguish PSCP from peritoneal carcinomatosis, a primary malignancy should be actively sought on imaging in all suspected patients. Peritoneal mesothelioma can be differentiated from PSCP on the basis of presence of lymphadenopathy and bone lytic lesions. In addition, mesothelioma has a different immunohistochemical profile with positivity for calretinin and keratin 5/6.

Multiple significant enlarged homogenously enhancing lymph nodes help suggest more a diagnosis of primary lymphoid disorder, especially in the presence of liver and splenic infiltration, rather than PSCP.

CONCLUSION

PSPC should always be considered in the differential diagnosis of a patient when metastatic ovarian cancer seems to be the diagnosis,
but the ovaries are normal on cross-sectional imaging. The other
differential diagnosis for this include malignant mesothelioma,
metastatic peritoneal carcinomatosis, peritoneal mesothelioma and
lymphomatosis, and should always be kept in mind.

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