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Case Report

PRIMARY RETROPERITONEAL SEROUS CYSTADENOMA WITH BORDLINE MALIGNANCY: A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

Background. Primary serous cystadenoma with bordline malignancy in the retroperitoneum is extremely rare entity. To our best knowledge, this is the first report in the world literature. The histogenesis is still unclear although several theories have been proposed. Such tumors are histologically similar to ovarian serous cystadenomas.

Case presentation. A 28-year-old woman presented with a six-month history of left-sided abdominal pain. Physical examination revealed a visible and a palpable oval solid mass in the left abdomen about 15 x15 cm in size. Computer tomography scan showed a homogenously hypoattenuating mass in the left retroperitoneal space. Laboratory blood studies as well as CEA, CA 19-9 were within normal limits. Laparotomy was performed through a left lateral paramedian incision. There were found a huge retroperitoneal cystic mass, measured 15 x 15 cm in size not adhered to adjacent organs. The tumor was unilocular, well-encapsulated by thin fibrous tissue, with a smooth surface. On opening the cyst contained a clear mucinous fluid. The uterus and both ovaries were normal. The cystic tumor was removed intact entirely and pathological examination revealed a primary serous ovarian-like cystadenoma with foci of borderline malignancy. The patient had an uneventful postoperative recovery and was discharged in good condition. There was no evidence of recurrence at her 8-years follow-up.

Conclusions. PRSC is extremely rare in surgical practice. Because its possess malignant potential, this rare entity should be considered in the differential diagnosis when a retroperitoneal cystic tumor is evaluated. The standard imaging methods – an abdominal US, CT scan, IMR confirm the presence of a cystic mass in the retroperitoneum, but have been of no aid in establishing an exact diagnosis. Only exploratory laparotomy with complete excision and a frozen section should approve the diagnosis. The prevention of cystic fluid spillage during surgery is important, especially when the pathology of the retroperitoneal cyst is unclear.

Key words: Retroperitoneal, primary, serous cystadenoma, bordline malignancy.

INTRODUCTION

Primary retroperitoneal serous cystadenoma (PRSC) is extremely rare clinical entity. To our knowledge only few cases have been described in the literature (1, 2, 3) and this is the first report of PRSC with bordline malignancy. The histogenesis is still unclear although several theories have been proposed. Such tumors are histologically similar to ovarian serous cystadenomas. We report a case of primary ovarian-like type PRSC with foci of borderline malignancy.

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CASE PRESENTATION

We reviewed the hospital record of a 28-year-old woman who presented with a six-month history of left-sided abdominal pain. She was detected a palpable mass in the left upper abdomen which was enlarged slowly over the past months. The past medical history revealed an uneventful pregnancy and delivery on time. Physical examination revealed a visible and a palpable oval solid mass in the left abdomen about 15 x15 cm in size. Laboratory blood studies as well as CEA, CA 19-9 were within normal limits. Ultrasonography demonstrated a 15-cm cystic mass in the region of the left abdomen. Computer tomography scan showed a homogenously hypoattenuating mass in the left retroperitoneal space, situated in the latero-caudal pararenal space **(Figure 1)**. For optimal exposure of the left retroperitoneum a transabdominal approach was performed through a left lateral paramedian incision. There were found a huge retroperitoneal cystic mass, measured 15 x 15 cm in size **(Figure 2).** The tumor was unilocular, well-encapsulated by thin fibrous tissue, with a smooth surface. On opening the cyst contained a clear mucinous fluid. We excluded attachment to any organ. The uterus and both ovaries were normal. The cystic tumor was removed intact entirely and pathological examination revealed a primary serous cystadenoma with borderline malignancy **(Figure 3)**.

The patient had an uneventful postoperative recovery and was discharged in good condition.

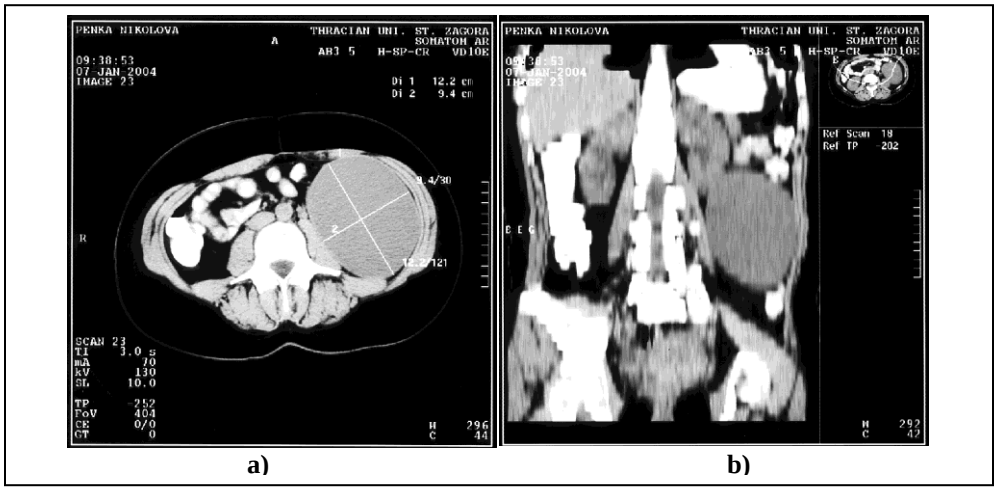

Figure 1. Abdominal CT showing a 12, 2 cm x 9, 4 cm homogenous, hypoattenuating, well-defined cystic mass, located latero-caudal pararenal in the left retroperitoneal space. a) axial and b) coronal reconstruction.

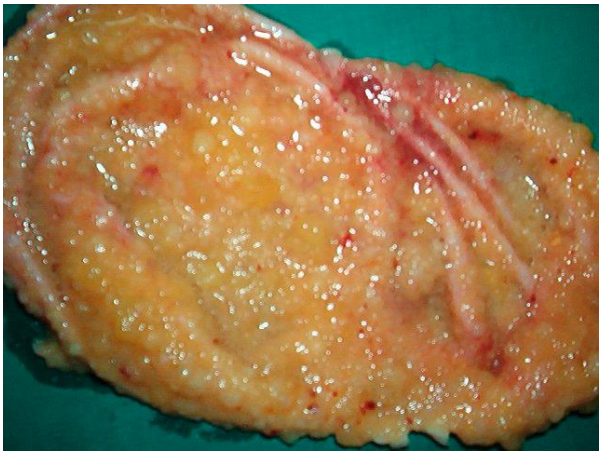

Figure 2. Photograph of the gross specimen shows cystic wall

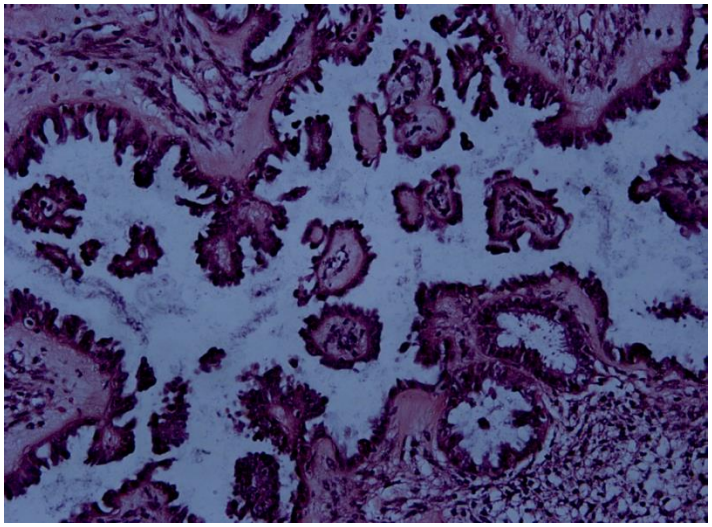

Figure 3. Serous borderline cystadenoma - complex, branching papillae with small papillary projections on the surface lined by epithelium showing cellular buds and nuclear stratification. Cells generally have moderate to abundant eosinophilic cytoplasm with round hyperchromatic nuclei and obvious nucleoli (hematoxylin and eosin stain, x40).

DISCUSSION

The primary epithelial neoplasms in the retroperitoneum remain extremely rare because of the non-existence of epithelial cells in this area. Grossly and histological, they resemble the epithelial tumors of the ovaries (ovarian-like tumors). They may be of serous or mucinous type and similar to their ovarian counterparts, can be further sub-divided into benign, with bordline malignancy and malignant. The unusual fact of these tumors is the retroperitoneal location in the presence of normal ovaries. The most common type of these epithelial neoplasms is primary retroperitoneal mucinous cystadenoma and their malignant variant - cystadenocarcinoma (4, 5, 6, 7, 8, and 9). Although several cases of primary serous cystadenocarcinoma have been reported (10, 11, 12, 13, 14), a careful search for PRSC by using the MEDLINE database revealed, as far as we aware, only three previously reported cases in the literature. According to most of the reported cases they are occurring almost exclusively in women with normal ovaries. Only six cases concerning male patients have been reported in the literature (15, 16, 17, 18, 19). According to G. Bifulco et al. the first description of this entity was given by Handfield in 1924 (4), but we found previously reported description of a retroperitoneal cystadenoma given by Staehlin (7). The clinical presentation usually is nonspecific and they are manifest with abdominal distention, discomfort depending on the size and location of the lesion. Correct preoperative diagnosis is sometimes impossible before surgical exploration and histological verification. Imaging studies such an abdominal US, CT scan and MRI may usually play an important role regarding the anatomic location, size and involvement of adjacent structures in the retroperitoneum, but failed to show its exact origin (12, 20). Laboratory evaluations such as carcinoembryonic antigen, alpha-fetoprotein, CA-19-9, CA-125, CA 15-3 rarely supported the diagnosis or follow-up (4, 3, 8, 11, 16). Most of the time these tumor markers were within normal limits except in rare cases. T. Sanefuji found an elevated concentration of a CA125 and CA19-9 in the cyst fluid (3). Motoyama et al. reported that measurement of CEA levels in the cystic fluid is useful in making the diagnosis (5).

Macroscopically and on microscopic examination the PRSC resemble serous cystic neoplasms of the ovary. The special feature in these cases is theirs localization in the retroperitoneal space without attachment to the ovary. Both neoplasms are usually a unilocular or a multilocular cystic mass with thin walls, contained mucinous fluid.

The histogenesis of primary retroperitoneal serous and mucinous cystic tumors remains unclear and several hypotheses have been proposed. Because they are grossly and histologically similar to serous and mucinous cystadenomas of the ovary mainly due to presence of ovarian-like stoma, one of the most accepted hypothesis is that these tumors arise from heterotopic ovarian tissue, or axillary ovary in the retroperitoneal space (9, 10). The supernumerary ovary has been considered the source for the development of benign or malign ovarian-like cystic mass in the retroperitoneum (11). However, ovarian tissue has been rarely identified in the cystic wall and with regard to the fact that these tumors appear in man weakens this hypothesis. The second convincing hypothesis proposed by Staehlin (7) that the retroperitoneal cyst arises from unabsorbed embryonic structures of the urogenital ridge. The third most widely accepted hypothesis suggests that these tumors originate from an invagination of multipotential mesothelial cells entrapped in the retroperitoneum. During the growing process these cells undergo a serous or mucinous metaplasia creating cystadenoma. These may then progress on to bordline and malignant serous or mucinous tumors (3, 7, 13, 16, 21, 23). According to Subramony et al. the presence of the flat to low cuboidal cells, positive for calretin, a mesothelial marker, support this origin (22).

Based on the review of cases reported in the literature, primary retroperitoneal epithelial tumors can be classified into 3 clinicopathologic types: benign retroperitoneal serous and mucinous cystadenoma, which are associated with no recurrences following total excision; serous and mucinous cystadenoma with bordline malignancy or low malignant potential – the lining epithelium contains foci of proliferative columnar epithelium in addition to the columnal epithelium; serous and mucinous cystadenocarcinoma, with high malignant potential – these patients usually have recurrent tumor and die from metastatic disease (9, 11). The patient we present was diagnosed with a primary retroperitoneal ovarian-like serous cystadenoma with foci of bordline malignancy.

Usually a surgical operation must be considered as the basic treatment of these neoplasms. Various opinions have been expressed about how extensive must the surgery be. In cases of benign cystadenoma everyone agrees on complete excision of the tumor (5, 8, 9, 18, 24). Some authors suggest laparoscopic resection of retroperitoneal cyst (25, 26). We are agreeing with them, especially if there is not any laceration of the cystadenoma's capsule and cystic fluid spillage during laparoscopy. An outstanding question is how extensive must the surgery be in cases of cystadenocarcinoma without involving an adjacent organs. Gotoh et al (9), Lee et al (29) suggest aggressive surgery of hysterectomy and bilateral salpingo-oophorectomy in addition to extirpation of retroperitoneal tumor, even though the uterus and ovaries are normal. We are agreed with de Leon et al. whose propose an aggressive adjuvant chemotherapy with hysterectomy and bilateral salpingo-oophorectomy only when involvement of these structures is found during the procedure (27).

In our patient, the PRSC with bordline malignancy was a well-defined cystic mass, did not infiltrate the adjacent organs. We performed complete evaluation of the abdominal and pelvic organs. The ovaries, uterus and other intra-abdominal structures were macroscopically normal. A total excision of the tumor, without hysterectomy and salpingo-oophorectomy was performed. There was no evidence of recurrence at her 8-years follow-up.

CONCLUSIONS

PRSC is extremely rare in surgical practice. Because its possess malignant potential, this rare entity should be considered in the differential diagnosis when a retroperitoneal cystic tumor is evaluated. The standard imaging methods – an abdominal US, CT scan, IMR confirm the presence of a cystic mass in the retroperitoneum, but have been of no aid in establishing an exact diagnosis. Only exploratory laparotomy with complete excision and a frozen section should approve the diagnosis. The prevention of cystic fluid spillage during surgery is important, especially when the pathology of the retroperitoneal cyst is unclear.

Abbreviations

PRSC - Primary retroperitoneal serous cystadenoma.

Competing interests
The authors declare that they have no competing interests.

Authors' contributions
AIP – reviewed literature and wrote the manuscript;
AMP – operated on the patient. Contributed to the concept and revision of the manuscript
TsAP - participated in the sequence alignment, researched sources for the references and drafted the manuscript;
V – Evaluated histopathology, contribution on images and critical revision of manuscript;
SPV – participated in the sequence alignment and helped to draft the manuscript.
GJJ– participated to searched literature and revision of manuscript.
All authors have read and approved the final manuscript.

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Consent
Written informed consent was obtained from the patient for publication of this case report.

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