

Primary Peritoneal Serous Psammocarcinoma: a Case Report

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ABSTRACT

Background: Psammocarcinomas (PCas) are rare epithelial tumors, usually originating in the ovaries or the peritoneum. These tumors are morphologically characterized by extensive psammomatous calcifications, invasiveness and low-grade cytological features.

Case report: We present the case of a 54-year-old woman who was referred to our department with an umbilical tumor and increasing abdominal girth. The patient had had an umbilical hernia for more than 20 years. The CA 125 level was normal. The CT scan showed small peritoneal nodules at the level of the Douglas pouch, including the posterior wall of the uterus, and the entire colon, as well as large nodules located on the caecum and the sigmoid colon. We performed partial enterectomy, total colectomy with ileo-rectal anastomosis, omentectomy, total hysterectomy and bilateral adnexectomy, pelvic peritonectomy of the Douglas pouch. Pathology findings were consistent with F.I.G.O. stage IIIC peritoneal PCa. The patient received adjuvant chemotherapy with Taxol and Carboplatin. To date, twelve months after surgery, the follow-up shows no evidence of disease.

Conclusion: Standardized treatment protocols are hindered by the rarity of the PCas. However, literature concludes that optimal debulking is mandatory, whereas the efficacy of adjuvant chemotherapy remains to be elucidated.

Key words: peritoneal psammocarcinoma – epithelial tumors – psammoma bodies.

Abbreviations: BRAF: proto-oncogene B-Raf; CA 125: cancer antigen 125; CEA: carcinoembryonic antigen; F.I.G.O.: International Federation of Gynecology and Obstetrics; LPD: leiomyomatosis peritonealis disseminata; PCa: psammocarcinoma.

INTRODUCTION

Psammocarcinoma (PCa) is a rare epithelial tumor, originating in the ovaries or the peritoneum, initially described by Kettle, while their pathological classification was established by Gilks et al. [1]. The characteristics of these tumors are the presence of numerous psammoma bodies, mild cellular atypia and invasiveness [2]. It seems that the clinical behavior of PCa resembles that of serous borderline tumors of the ovaries or peritoneum, rather than of typical invasive papillary serous carcinomas, with a prolonged

clinical course and relatively favorable prognosis [3, 4]. Peritoneal PCa is even rarer than ovarian PCa, with only 23 cases described in the English-language literature [5]. We present the case of a 54-year-old woman with primary peritoneal PCa, suspected by CT-scan and confirmed by pathologic findings after surgical biopsy.

CASE REPORT

A 54-year-old postmenopausal woman was referred to our hospital with a 14 cm umbilical tumor, increasing abdominal girth in the past three years and progressive weakness. She had not received any hormone therapy and had not experienced postmenopausal bleeding. Her past medical history included umbilical hernia for more than 20 years, which grew in size and became hard on palpation in the last two years. Physical examination revealed stable vital signs, and a protuberant non-tender abdomen, due to the presence of a firm umbilical tumor. Pelvic examination revealed no abnormalities. Pap test, serum

level of CA125 and CEA were normal. Computed tomography (CT) scan showed small nodules in the Douglas pouch, and on the surface of the colon, large nodules located on the omentum, caecum (3.5/2.9 cm), and sigmoid colon (3/2.6 cm) and a 14 cm umbilical tumor, invading the ileum. All tumors were high-density tumors. Calcifications on the caecum, a large, calcified umbilical tumor, and fine calcifications on the great omentum are shown in Fig. 1. Upper digestive endoscopy and colonoscopy were normal.

To establish the diagnosis, we performed a surgical biopsy under local anesthesia. Pathological evaluation of the biopsy sample revealed a serous carcinoma, with multiple psammoma bodies, probably arising from the peritoneum or the ovaries. During open surgery, the umbilical tumor was removed in block with a segment of distal ileum, and was followed by total colectomy with ileo-rectal anastomosis, omentectomy, total hysterectomy, bilateral adnexectomy, and local peritonectomy of the Douglas pouch. The patient was optimally debulked with no gross tumor remaining. Pathology findings were consistent with the International Federation of Gynecology and Obstetrics (FIGO) stage IIIC peritoneal PCa. The great omentum was entirely replaced by invasive tumor implants. The common feature of all these tumors was the presence of numerous psammoma calcifications (psammoma bodies) (Fig. 2). Four out of the 33 retroperitoneal lymph nodes had psammoma calcifications invading the lymph nodes (Fig. 3). The patient received post-operative adjuvant systemic chemotherapy with Taxol 175 mg/m² and Cisplatin 75 mg/m², six cycles every three weeks, without any side effects. Ten months after surgery she is alive and well, without local or distant metastases.

DISCUSSION

Psammocarcinoma is a rare form of invasive papillary serous carcinoma associated with extensive psammoma body formation [6, 7]. Psammoma bodies are calcified spherules measuring 50-100 µm in diameter with concentric laminations. They are thought to be formed secondary to the accumulation of hydroxyapatite in cells that undergo dystrophic calcifications related to cellular degeneration [5, 7].



Fig. 1. High-density tumors on a native CT-scan in the umbilicus, caecum and great omentum (white arrows).

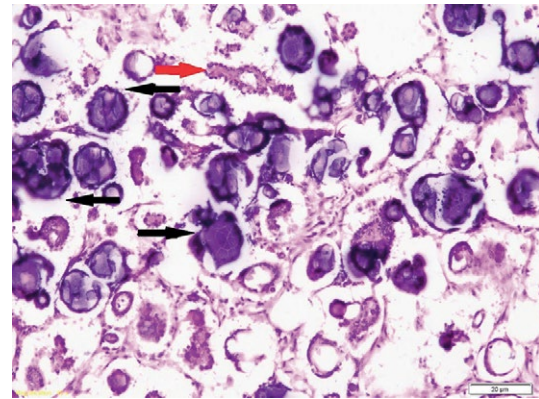


Fig. 2. Histological aspect of the multiple psammoma bodies (black arrow) and epithelial cells (red arrow) (H&E x20).

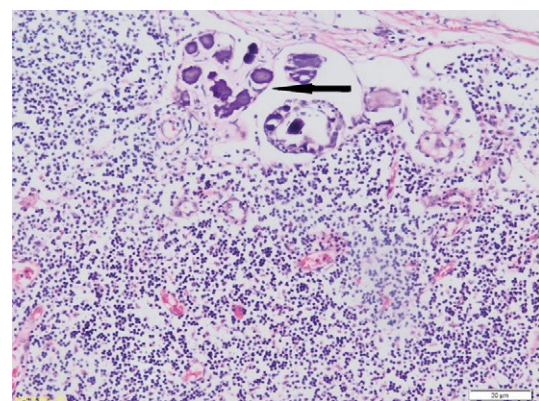


Fig. 3. Histological aspect of a lymph node invaded by psammoma bodies (black arrow) (H&E x10).

They are found commonly in certain human neoplasms, such as thyroid, meningeal, ovarian and also in gastrointestinal tumors (duodenal carcinoid, and gastric adenocarcinoma) [8-11]. Pathologic differential diagnosis of PCa should be done with other serous epithelial tumors. Serous adenocarcinomas usually have high-grade atypia, and behave aggressively, whether or not psammoma bodies are present. On the other hand, PCa differs from well-differentiated serous adenocarcinomas due to the formation of numerous psammoma bodies, in at least 75% of the papillae [12]. Differential diagnosis of PCa should be made also with calcified leiomyomatosis peritonealis disseminata (LPD), a rare, particular type of leiomyomatosis with unclear pathogenesis, usually affecting females of reproductive age with a surgical history of laparoscopic myomectomy for uterine fibroma, and in whom laparoscopic power morcellation was used [13]. Our patient was a 54-year-old postmenopausal woman, without a history of a laparoscopic myomectomy, thus the diagnosis of LPD was excluded.

In 1990, Gilks et al. defined four specific histologic diagnostic criteria for PCa: 1) destructive invasion of the ovarian stroma or of the intraperitoneal viscera and peritoneum; 2) no more than moderate cytology atypia; 3) no areas of solid epithelial proliferation except for occasional nests, with no more than 15 cells in diameter, and 4) at least 75% of the papillae associated with or completely replaced by psammoma body formation [2, 14]. In 1994, Chen et al. expanded Gilks's

criteria for diagnosis of peritoneal PCa to include either invasions of the intraperitoneal viscera or an invasive pattern in the peritoneum [15]. Unlike peritoneal PCa, serous PCa of the ovary is a rare ovarian neoplasm characterized by extensive psammoma body formation, peritoneal or intraperitoneal visceral infiltration, moderate cytologic atypia, and the most important feature, ovarian stromal invasion [2, 16, 18]. According to Gilks's criteria, extended by Chen, our case was designated as a primary peritoneal PCa. The diagnosis of PCa is mainly dependant on histopathological findings.

The clinical presentation of primary peritoneal PCa is similar to that of FIGO III ovarian and peritoneal serous borderline tumors or carcinomas, with nonspecific signs and symptoms of increasing abdominal girth, discomfort or both [6]. In some cases, clinical presentation has included elevated serum CA-125 levels. However, as with serous borderline tumors, elevation of CA-125 if it occurs, is moderate [4, 6]. High level of serum CA125 may show potential for aggressive behavior [13]. The largest series of PCa patients was published by Weir et al. in 1998. In this report, patients had an average age of 40 and primarily presented with abdominal pain or an abdominal mass. They also indicated that PCa has similar clinical and pathologic characteristics with low-grade serous carcinoma and serous borderline tumors [16]. It seems that the clinical behavior of peritoneal PCa, like that of ovarian PCa, resembles more closely to that of serous borderline tumors rather than that of typical invasive papillary serous carcinomas [4, 7].

Genes of the RAF family encode kinases that are regulated by RAS. Activating mutations in BRAF have been identified in ovarian tumors and other malignancies such as melanomas, thyroid or colorectal cancers. Sieben et al. showed that BRAF mutations in ovarian tumors occur exclusively in low-grade serous neoplasms, which include PCa [17]. We did not evaluate the tumors for BRAF mutation in our patient.

There is no standardized management of peritoneal PCa, due to its rarity [6, 7, 12, 18, 19]. However, most authors recommend optimal surgical debulking of the tumor and adjuvant chemotherapy [2, 6, 12, 20]. The mainstay of the treatment is maximal surgical debulking, including omentectomy, total abdominal hysterectomy and bilateral salpingo-oophorectomy [6]. Conservative surgery is an acceptable option only in young women diagnosed with PCa [16]. Munkarah et al. consider that responsiveness of ovarian and peritoneal PCa to chemotherapy and benefit of its treatment postoperatively have not been established and are subjects of debate [16]. Molpus et al. reported a case with recurrent peritoneal PCa with complete response to tamoxifen therapy [7]. Weir et al. also administered tamoxifen in their patients with recurrent disease, although survival appears to be maximized by surgical removal alone [15].

In our patient, optimal debulking was performed, followed by post-operative adjuvant systemic chemotherapy with Taxol and Cisplatin. The treatment was well tolerated by the patient without any side effects.

CONCLUSIONS

The peritoneal PCa is a rare serous tumor, characterized by low-grade cytological features, invasiveness and the formation

of numerous psammoma bodies. Biological behavior of these tumors is less aggressive than that of serous peritoneal carcinomas. The mainstay of the treatment is represented by optimal surgical debulking. Regarding postoperative treatment, there is no definite evidence that adjuvant chemotherapy improves survival.

Conflicts of interest: None to declare.

Authors' contribution: R.T. and V.M. designed the report and critically revised the manuscript; A.C. searched literature data; R.T., A.Z. and V.M. analyzed and interpreted data; R.S. and I.S. drafted the manuscript.

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