Psammocarcinoma is a rare variant of serous carcinoma arising either from ovary or peritoneum, characterized by massive psammoma body formation, low-grade of cytologic differentiation and invasiveness. Its clinical behavior is similar to the serous borderline tumors, whose prognosis is significantly better compared to invasive forms, with a 5-year survival in stage I greater than 95%. A typical feature of borderline ovary tumors is the presence, in more than 30% of cases, of borderline peritoneal implants similar to primary ovarian cancer or of invasive forms.

We report a case of a 44-years-old woman who referred to our clinic for mesosigmoid mass, accidentally discovered by ultrasonography. Sigmoidectomy with fertility sparing surgery was performed in September 2010. The mass was histologically characterized by many psammoma bodies and low grade cytological features with diagnosis of psammocarcinoma of mesosigma. One year after the primary surgery, the patient showed with left adnexial mass; optimal debulking surgery was performed including omentectomy, total abdominal hysterectomy, bilateral adnexectomy and appendicectomy. The patient did not receive any adjuvant chemotherapy and to date she is alive and with no evidence of disease.

The conclusion is that psammocarcinoma is a very rare tumor that behaves less aggressively than typical serous carcinoma, the mainstay of treatment is surgical debulking, with fertility sparing surgery as possible option in young patients with ovaries macroscopically free of disease.

KEY WORDS: Borderline neoplasia, Psammocarcinoma, Rare tumor.

Introduction

Borderline tumors are from 10 to 20% of all epithelial ovary tumors and, differently from the invasive forms, they have the tendency to show up at an earlier stage and at a younger age. The prognosis is remarkably better than the invasive forms, with a survival rate at 5 years of I stage (which includes 75-85% of such tumors) above 95% \(^1\). These are tumors mainly made of serose and mucinous forms, while endometrioid forms as well as clear cells and Brenner ones are rare \(^2\). Serose borderline tumor is bilateral in 25-50% of cases; this percentage drops for the other histological type. Psammocarcinoma is a rare subtype of serous adenocarcinoma that can arise from the ovaries or peritoneum (PSPP) \(^1\). The PSPP is even more rare than its ovarian counterpart \(^2\). To our knowledge there are nearly 25 cases reported in literature \(^3,6\). Histologically they are characterized by the presence of psammomatous calcifications.
(psammoma bodies), invasiveness and low grade cytological features \(^1,2\), as the borderlines tumors. A typical feature of borderline ovary tumors is the presence, in more than 30% of cases, of borderline peritoneal implants similar to primary ovarian cancer or of invasive ones. If such implants are metastases of the primary cancer or are the occurring of peritoneal multifocal lesions is not yet clearly understood, even if literature data state the multifocal origin as the most likely one. These tumors usually have a painless course and carry a more favorable prognosis than the invasive serous adenocarcinoma, with progression and distant metastasis in only a small minority of cases \(^1,3,4,6\). Due to their rarity, there is no established standard therapy for these tumors: the mainstay of treatment is maximal surgical debulking, the role of adjuvant chemotherapy on improving survival requires further evaluations \(^2,6\). We describe a case of mesosigmoid psamnocarcinoma.

**Case report**

In September 2009, a 44 years old women (gravida 2) came to us for a multiseptated mass measuring 60x55x90 mm in Douglas pouch, detected during a pelvic ultrasound performed for age-related screening. The patient was totally asymptomatic, CA 125 levels were found normal. Pelvic RM, performed in September 2010 (Fig. 1), revealed that the mass had a complex structure for the presence of a solid peripheral portion of low signal and a central portion hyper-intense in T1 gated sequences, probably hematic, multiseptated. The mass appeared poorly dissociable from the wall side of the sigmoid colon, so determining compression and dislocation of it. At the end of September 2010 the patient underwent surgery and was found to have a solid mass in mesosigma infiltrating the sigmoid wall at mesenteric side; the ovaries were macroscopically normal without any involvement (Fig. 2). Due to this finding, and to the young age of the patient who wished to preserve her fertility, it was performed a sparing fertility surgery, with segmental sigmoidectomy and subsequent colo-rectal end to end anastomosis. No visible residual tumor was left. The histologic exam revealed a serous papillary psammocarcinoma characterized by massive psammoma bodies formation. Mitotic index <1/10 HPF, the neoplasia invaded the wall of sigmoid colon, infiltrating the muscularis propria layer. CK7+, CA125+, CK20-, CEA-. Post operative recovery was uneventful. Following the surgery the patient did not receive chemotherapy because of low mitotic index, the low cytologic grade of tumor and the effectiveness of surgery. In August 2011, during the follow up, a PET scan (Fig. 3) revealed an high radionuclide captation (SUV 10,6 max) of probable uterine relevance. A transvaginal ultrasound, performed in September 2011, showed a complex mass, poorly vasocarlarized measuring 30 mm, at the level of left ovary, with diagnosis of recurrent disease. In October 2011, the patient underwent second laparotomy that showed a left para-ovaric mass without any involvement in peritoneum. She was treated with total hysterectomy, bilateral salpingo-oophorectomy, appendicectomy, omentectomy and pelvic lymphadenectomy with peritoneal washing. Microscopic findings were consistent to a psammocarcinoma in F.I.G.O. stage IA G1, the omentum and all the lymphonodes removed were disease free, with only involvement of left ovary; the cytopatologic examination showed only inflammatory cells. The post-operative course was regular with discharge from our clinic in day
seventh. The patient did not receive adjuvant chemotherapy because of the stage (IA G1). At the time she is alive without evidence of disease.

Discussion

First described by Kettle in 1916, psammocarcinoma is a rare form of low-grade serous carcinoma (low proliferative activity with Ki67 and diploid DNA) arising either from ovary or peritoneum, histologically characterized by the pervasive presence of psammoma bodies measuring 5-10 microm in diameter, formed by accumulation of hydroxyapatite in degenerated cells that suffered dystrophic calcification. The psammoma bodies may be found in various neoplastic and non-neoplastic lesions like papillary thyroid carcinoma, cranial meningioma and ovarian serous papillary carcinoma: tumors with low malignant potential and less aggressive clinical course with a milder biological behavior; however prognosis significance of psammoma bodies is still unclear due to different study results.

Gilks et al. in 1990 proposed histologic criteria for diagnosis of psammocarcinoma:
- Destructive invasion of the ovarian stroma or intraperitoneal viscera;
- No more than moderate cytologic atypia;
- No areas of solid epithelial proliferation except for occasional nesting no more than 15 cells in diameter;
- At least 75% of papillae are associated or completely replaced by psammoma body formation.

According to these criteria and the revised criteria established by Chen, our case was defined as a primary psammocarcinoma (PSPP) of mesosigma; bilateral ovaries at time of first diagnosis were grossly normal. A total of about 25 cases of peritoneal and 20 cases of ovarian psammocarcinoma are found in literature; the usual localization in the mesosigma with invasion of sigmoid muscolaris and serosa was never reported before.

Psammocarcinoma has been suggested to have more favorable prognosis compared to invasive serous adenocarcinoma ovarian counterparts; however Weir et al. reported the data regarding the evaluation of clinical outcome in peritoneal serous psammocarcinoma and peritoneal serous borderline tumors showing no difference in term of survival. As in our case psammocarzina may have recurrence, in other cases may show a more aggressive behavior with metastases and elevated CA-125.

The clinical presentation of peritoneal psammocarcinoma is characterized by non specific signs and symptoms of increasing abdominal girth, discomfort or both in most cases, nausea and vomiting, heavy menstrual bleeding, while some were asymptomatic or accidentally detected as our case. CA 125 levels was usually detected high in psammocarcinoma of ovary but normal in PSPP; however as with serous borderline tumors, increase of CA 125, if it occurs, is moderate.

Psammoma bodies are rarely found in cervicovaginal Pap smears; nevertheless, the diagnosis of intrabdominal psammocarcinoma should seriously be taken into account in any patient with this findings. Rarely the presence of endometriosis, endosalpingiosis and mullerian inclusion cysts has been documented in PSPP and in ovarian psammocarcinoma.

Conclusion

Because of the rarity of this tumor, there is no standard in the management of these patients; the therapy regimen for the psammocarcinoma differs among the institutions, in the most of cases the treatment has been tailored to the patient situation: the protocols include usually total abdominal hysterectomy with bilateral salpingo-ooforectomy, lymphadenectomy (always pelvic and retroperitoneal only in cases of large implants >2 cm), omentectomy, appendicectomy and maximal tumor debulking. Conservative surgery (fertility sparing approach), as in our case, is an acceptable option in young women, also in women who desire to preserve their fertility, in all cases when bilateral ovaries were macroscopically normal without any involvement.

The rarity of psammocarcinoma has made assessment of adjuvant systemic chemotherapy in the treatment of this disease very difficult; in few cases with PSPP who received adjuvant chemotherapy there was no documentation of an objective response; in patients with papillary serous neoplasm of low malignant potential (border-line tumors), tumor responsiveness to chemotherapy and the benefits of such treatment continue to be a subject of debate. In patients with papillary serous neoplasm of low malignant potential (border-line tumors), tumor responsiveness to chemotherapy and the benefits of such treatment continue to be a subject of debate. There are no randomized trials that have evaluated the efficacy of adju-

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Riassunto

Lo psammocarcinoma è una rara variante di carcinoma sieroso che origina dalle ovaie o dal peritoneo, caratterizzata da un elevato numero di corpi psammomatosi, basso grado di differenziazione citologica ed invasività. Mostra un comportamento clinico simile a quello dei tumori sierosi border-line. Riportiamo il caso di una donna di 44 anni, giunta alla nostra osservazione per una massa pelvica, scoperta per caso nel corso di un esame ecografico. Alla laparotomia esplorativa, effettuata nel settembre del 2010, la massa risultò essere di pertinenza del mesosigma, con infiltrazione della parete colica; indenni apparivano le ovaie e l’utero. Fu eseguito un intervento di sigmoidectomia. La neoformazione all’esame istologico risultò essere uno psammocarcinoma del mesosigma. Un anno dopo l’intervento, la paziente pratica ecografia trans-vaginale di follow-up, che evidenziava una massa a carico dell’ovaio di sinistra; veniva sottoposta ad intervento chirurgico di isterectomia totale, annessiectomia bilaterale, linfodenectomia pelvica, appendicectomia ed omentectomy. L’esame istologico definitivo evidenziava uno psammocarcinoma ovarico sinistro, stadio IA G1, sec. F.I.G.O., liberi da malattia l’omento ed i linfonodi isolati. La paziente, in base allo stadio anatomico – patologico, non è stata sottoposta ad alcuna chemioterapia adiuvante ed attualmente gode di buona salute, inserita in uno stretto protocollo di follow-up diagnostico – strumentale.

In conclusione, da una accurata revisione della letteratura, questo sembra essere il primo caso di localizzazione dello psammocarcinoma a livello del mesosigma. Tali neoplasie, vanno considerate come tumori sierosi border-line a prognosi favorevole, ma comunque in grado di dare recidive locali e metastasi a distanza. Il trattamento chirurgico rappresenta l’opzione terapeutica di prima scelta, la possibilità di una chemioterapia va valutata in base allo stadio della neoplasia, c’è scarsa evidenza che una terapia adiuvante possa migliorare la prognosi già buona di questi tumori rari.

References


