Primary Retroperitoneal Mucinous Cystadenoma with Sarcomatous Carcinoma in Mural Nodule

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Abstract Primary retroperitoneal mucinous cystic tumors are extremely rare neoplasms that usually occur in women. Their histogenesis has not been clarified. Their low incidence has meant that it difficult to define therapeutic strategies or prognosis, or to record survival data. We describe a 35 year-old female with a retroperitoneal mucinous cystadenoma presenting a mural nodule with foci of sarcomatoid carcinoma. The entire tumor was surgically removed. After surgery, the patient received six cycles of carboplatin and paclitaxel. After 30 months of follow up she has shown no signs of recurrence. We review the etiology, diagnosis and therapeutic options in these extremely rare tumors.

Keywords: retroperitoneal, tumor, surgery, mucinous, cystic, sarcomatoid, review

Introduction

Primary retroperitoneal mucinous cystic tumors (PRMC) are extremely rare neoplasms usually occurring in women. Their histogenesis has not been clarified [1-11]. The few reports to date have been mostly isolated cases or case reviews [1-11], and very little information is available for defining therapeutic strategies and determining prognosis and survival. We report a patient with a mucinous cystadenoma presenting a mural nodule with foci of sarcomatoid carcinoma.

Case Report

Thirty-five year old female, without relevant medical history, presented at our Outpatient Clinic with abdominal pain and self-palpation of a mass in the left flank. Physical examination identified a non-fixed mass occupying the entire left flank. The analyses performed, including CEA and CA19-9, were all normal. The ultrasound showed a lesion of 14cm of maximum diameter with a small solid nodule of 2cm. The CT (Figure 1) and MRI revealed a cystic mass of 14 x 10 x 9 cm in the left flank with a thin wall. A well-defined nodule was seen in the inner wall of the mass. The tumor was located above the left kidney and psoas, displacing the left colon (Figure 1). No locoregional lymphadenopathy or free fluid was seen. Laparotomy was performed and the cystic lesion was removed intact (Figure 2). Macroscopically, the specimen was a cystic mass weighing 697.4g with a smooth and pearly wall (Figure 3). The cyst fluid was brown and mucoid. The inner surface was smooth; the nodule, measuring around 2cm, was brown, solid, homogeneous, soft and elastic, with yellowish necrotic areas. Histological examination showed an inner wall lined internally by a conjunctival epithelium without papillary projections and abundant mucoid material in the cytoplasm [1]. No proliferation, invasion, or atypia was observed in the inner wall of the cyst. The nodule presented a homogeneous and dense cellular proliferation with superficial necrosis composed of elongated pleomorphic cells, often multinucleated, with a sarcomatous appearance. The nodule stroma was fusocellular and presented necrosis and inflammatory lymphoid cells. The nodule showed no wall infiltration or extracapsular extension. The definitive histological diagnosis was mucinous cystadenoma with sarcomatoid carcinoma nodule of 20mm. Immunohistochemistry study was positive for high and low molecular weight cytokeratin (CK 7, CK17 and CK20) in the cystic wall and also in the nodule. CD68 was weakly positive. p53 was positive in most of the atypical nodule cells. Results were negative for CD34, s100, alpha-actin, actin, vimentin, estrogen and progesterone receptors. The rate of cell proliferation was low except in the nodular area. It was decided to start treatment with chemotherapy (six cycles of carboplatin and paclitaxel). After 30 months’ follow-up, the patient shows no signs of recurrence.

Retroperitoneal mucinous cystic tumors (PRMC) are extremely rare [1,5,6,7]. No more than sixty cases have been described to date. The first report was by Handfield-Jones in 1924 [10]. PRMC are usually seen in women in the fourth to sixth decade of life, with a mean age around 40 years [5,10]. The few men with PRMC tend to be older (60-80 years) [5,11]. PRMC usually are unilateral and large (mean: 15cm) (1.7). Their gross appearance varies: they may be unilocular or multilocular, with or without nodules, or they may be solid-cystic lesions [1].

PRMC are classified into three types according to their pathological features. Mucinous cystadenoma, the benign variant, is the most common. Borderline mucinous...
cystadenoma presents proliferative foci of columnar epithelium and has a low malignant potential. Mucinous cystadenocarcinoma may metastasize and/or recur (2,3,4,7,8,10,11). In immunohistochemical studies, PRMC are always positive for cytokeratins CK7 and CK17 and in some cases for CK20 [1,10]. A very important prognostic factor in PRMC is the presence or absence of mural nodules. These nodules can be malignant (anaplastic carcinoma, sarcomatoid carcinoma or true sarcoma), or benign. If these benign nodules present sarcomatoid appearance they are called pseudosarcomatous or sarcoma-like nodules [7].

Our extensive literature search found only descriptions of cystadenocarcinomas [1,7], and so, to our knowledge, this is the first report of a benign lesion (cystadenoma) showing a malignant mural nodule.

Figure 1. CT: Primary retroperitoneal mucinous cystadenoma. (*) nodule

Figure 2. Operative field

Figure 3. Surgical specimen

Several hypotheses have been proposed regarding the origin of PRMC. It has been suggested that they are teratoma-like lesions whose columnar epithelium has become the predominant cell, or ectopic ovarian tissue or supernumerary ovaries (though few PRMC present traces of ovarian tissue, and immunohistochemical staining for estrogen and progesterone is usually negative) or embryological remnants of the urogenital system. The most widely accepted theory is coelomic metaplasia: during embryonic development invaginations of retroperitoneal mesothelium occur in the retroperitoneal space, forming inclusion cysts which undergo mucinous metaplasia [1-7,10]. The positivity for EMA and the selective positivity for vimentin support this theory [1-7,10].

PRMC may be asymptomatic and may only be diagnosed incidentally on imaging tests performed for other medical reasons [10,11]. When symptoms are present, they tend to be pain or bloating due to tumor compression [1,2,5,7,10]. It is not uncommon for patients to self-palpate the lesion, as occurred in our case [10].

CT and MRI are the most cost-effective imaging methods for the diagnosis of retroperitoneal cystic lesions [2,6,7,11]. In abdominal CT, PRMC are seen as homogeneous unilocular cystic masses [6]. We stress that mural nodules may be seen in CT and MRI. Differential diagnosis of PRMC includes cystic mesothelioma, cystic lymphangioma, mesothelial cysts, enteric duplication cysts, lymphocele, cystic teratoma and pancreatic pseudocyst [3,4,6,10]. Performing a preoperative FNAC is controversial because even though it may help preoperative diagnosis, it does not always differentiate between malignant and benign tumors and may cause neoplastic spreading in malignant cases [6]. In agreement with other authors [5], we argue against the use of FNAC.

Serum tumor markers (CA19-9, CA125 and alpha-fetoprotein) are relatively unhelpful because their rates are usually normal. However, some patients may present significant increases [2,5,10]. Elevated intracystic levels of CEA and CA19-9 have also been observed [3].

The treatment of PRMC is complete surgical excision of the lesion without opening [4,5,6,10]. Classically, surgery was performed by laparotomy, although one case performed via laparoscopic approach has been reported [8]. We decided to perform laparotomy due to the existence of a potentially malignant mural nodule and due to our concern regarding a possible iatrogenic opening of the cyst and the spread of its contents into the abdomen.

In women with the cystadenocarcinoma variant of PRMC, some authors propose hysterectomy and bilateral oophorectomy due to its possible gynecological origin. However, the postoperative histological studies of these surgical specimens have shown no alterations [1,2,5,9,11]. Currently, it is generally accepted that if the uterus and ovaries are macroscopically normal, as in our patient, removal is not justified.

Adjuvant chemotherapy is recommended in malignant cases, especially if there is spontaneous or intraoperative rupture of the lesion, sarcoma or anaplastic nodules, or invasion of adjacent structures [2,5]. In the series reported by Rome et al, a patient who suffered a cystadenocarcinoma with sarcomatoid nodule was treated with ifosfamide, cisplatin and paclitaxel [1]. Some authors recommend adapting the protocols used in mucinous ovarian tumors [2].
Patients with cystadenoma or borderline cystadenoma and complete removal of the lesion have an excellent prognosis. However, the presence of a sarcoma or anaplastic sarcomatoid carcinoma nodule has a negative impact on prognosis; metastases are frequent in these patients and survival is poor [1].

References