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Case report

PRIMARY RETROPERITONEAL TUMOR: MUCINOUS CYSTOADENOCARCINOMA

MARIANA TOFFOLO PASQUINI[1], LUCÍA ARAGONE[1], VICTORIA SCASSO REBDZA[1], WALTER NARDI[2], MARÍA TOSCANO[3], SERGIO QUILDRIAN[2]

[1]Departamento de Cirugía General, [2]Unidad de Sarcomas y Melanomas, Departamento de Cirugía General, [3]Departamento de Anatomía Patológica, Hospital Británico de Buenos Aires, Argentina

Postal address: Mariana Toffolo Pasquini, Hospital Británico de Buenos Aires, Perdriel 74, 1280 Buenos Aires, Argentina
E-mail: marianatoffolo@gmail.com
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Abstract

Primary retroperitoneal mucinous cystadenocarcinomas (PRMCs) are extremely rare tumors with limited understanding of their pathogenesis and biological behavior.

We describe a case of a 50-year-old female patient who underwent surgical treatment. The patient had a history of previous surgeries for mesenteric mucinous cystadenoma, without evidence of recurrence. During routine abdominal ultrasound a new tumor was found. An abdomen magnetic resonance imaging was done and confirmed the presence of a cystic lesion in the right iliac fossa.

After discussion in multidisciplinary committee, surgical complete resection of the tumor, along with bilateral adnexectomy, was performed successfully. Histopathological examination revealed a mucinous adenocarcinoma adjacent to a mucinous cystadenoma. Immunohistochemical analysis supported the diagnosis of a primary retroperitoneal lesion. The patient had an uneventful recovery and has remained disease-free during the two-year postoperative follow-up.

PRMCs are challenging to diagnose preoperatively due to nonspecific symptoms. Surgical excision is the mainstay of treatment. The long-term prognosis and optimal therapeutic strategies require further investigation.

Key words: primary cystadenocarcinoma, mucinous tumor, retroperitoneum, cystic tumor

Resumen

Tumor primario retroperitoneal: cistoadenocarcinoma mucinoso

Los cistoadenocarcinomas mucinosos primarios retroperitoneales (CMPR) son tumores extremadamente raros con una comprensión limitada de su patogénesis y comportamiento biológico.

Describimos el caso de una mujer de 50 años sometida a tratamiento quirúrgico. La paciente tenía antecedentes de cirugías previas por cistodenoma mucinoso mesentérico, sin evidencia de recurrencia. Durante una ecografía abdominal de rutina se encontró un nuevo tumor. Se realizó una resonancia magnética abdomen que confirmó la presencia de una lesión quística en la fosa ilíaca derecha.

Luego de discutir el caso en el comité multidisciplinario, se realizó con éxito la resección quirúrgica completa del tumor, junto con la anexectomía bilateral. El examen histopatológico reveló un adenocarcinoma mucinoso adyacente a un cistodenoma mucinoso. El análisis inmunohistoquímico apoyó el diagnóstico de lesión primaria retroperitoneal. La paciente tuvo una buena recuperación y permaneció libre de enfermedad durante dos años de seguimiento postoperatorio.

Los CMPR son difíciles de diagnosticar debido a que presentan síntomas inespecíficos. La escisión quirúrgica es la base del tratamiento. El pronóstico a largo plazo

y las estrategias terapéuticas óptimas requieren más investigación.

Palabras clave: cistadenocarcinoma primario, tumor mucinoso, retroperitoneo, tumor quístico

Mucinous adenocarcinomas are relatively frequent tumors. However, primary retroperitoneal mucinous cystadenocarcinomas (PRMCs) are extremely rare[1,2]. While the vast majority of documented cases involve female patients, some cases have been reported in male patients. Given this neoplasm´s low incidence, its pathogenesis and biological behavior are not well understood.

We report one case of PRMC in a patient that underwent surgical treatment. We report this case according to the Updating Consensus Surgical CAse REport (SCARE) 2020 guidelines[3].

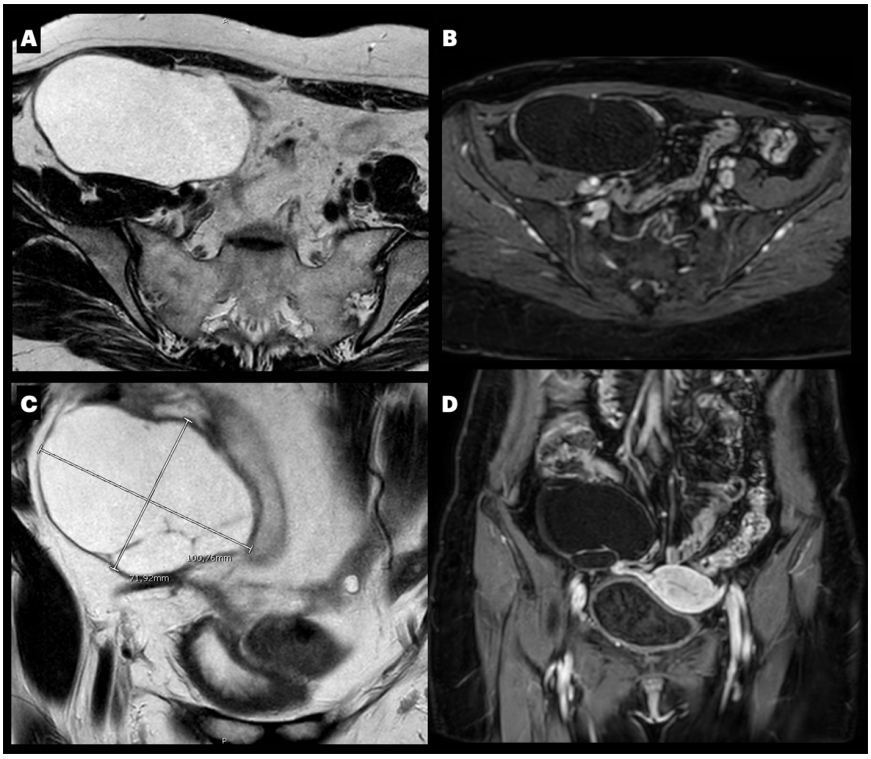
Clinical case

A 50-year-old female patient was referred to our institution after abnormal findings in a routine abdominal ultrasound. Her medical history was significant for IgA nephropathy, dyslipidemia, laparoscopic right ovarian cystectomy and laparoscopic resection of a mesenteric cyst performed at another institution, 21 years prior to consultation at our center. The pathology reported mucinous cystadenoma. She continued follow-up for 20 years without evidence of recurrence.

The patient was in good general condition. Physical examination revealed a painless abdomen without any palpable masses. The control abdominal ultrasound reveled a cyst in the right iliac fossa. To characterize the lesion, an abdomen magnetic resonance imaging (MRI) was done, showing a cystic-looking image measuring 101 × 72 × 74 mm, with fine septa inside, located in the right iliac fossa. It presented parietal and septal enhancement with intravenous contrast and a 7 × 6 mm nodular image on its posterior wall, as well as bilateral ovarian cystic images (Fig. 1). A transvaginal ultrasound was performed where a 34x12 mm fluid-containing thin-walled tubular-looking image was observed in the right adnexal region with echogenic nodular formation at its distal end of 8x4 mm. Tumor markers, including CA 19-9 (<3 u/ml), CA 125 (<3 u/ml) and CEA (1.9 ng/ml), were within normal limits.

After discussion in multidisciplinary committee, surgical resection of the cyst associated with bilateral adnexectomy was decided due to suspicion of adnexal blastoma.

Figura 1 | Abdomen MRI: Cystic-looking image measuring 101x72x74 mm, with fine septa inside, located in the right iliac fossa, with enhancement with IV-contrast of the cyst wall A: Axial view, T2 sequence. B: Axial view, T1 sequence, with IV constrast. C: Coronal view, T2 sequence, showing measurement of the cyst. D: Coronal view, T1 sequence, with IV constrast



The patient underwent complete surgical excision of the tumor via a laparotomic approach due to its size. A retroperitoneal mesenteric cyst of approximately 8 cm of diameter was identified (Fig.2 A). Its complete resection was achieved without rupture of the tumor (Fig.2 B). In addition, bilateral adnexectomy was performed. Both surgical pieces were sent for histopathological examination, which revealed a serous cystadenoma at the annexes associated with a paratubal serous cystadenofibroma, and a moderately differentiated mucinous adenocarcinoma, adjacent to mucinous cystadenoma. The following immunohistochemical markers were positive: CK7, CK 19, CK20 (focal), CDX2 (focal), receptor estrogénico (focal and isolated to capsule´s nucleus) and PAX 8 (focal and isolated to capsule´s nucleus). TTF1 was negative and P53 showed no overexpression (Fig. 2 C-F). After ruling out the existence of a mucinous lesion of the ovary, we concluded that the histological characteristics and the immunological profile are consistent with a primary lesion of the retroperitoneum.

The patient had an uneventful recovery and was discharged on postoperative day three. Outpatient controls at first postoperative week and every 6 months were unremarkable.

Currently she is undergoing 2 postoperative years without evidence of recurrence by physical examination or imaging, with abdomen and pelvis MRI.

With the final result of the pathology, it was discussed in multidisciplinary committee and it was decided to continue outpatient controls with physical examination and MRI every 6 months.

The patient had signed the corresponding informed consent form.

Discussion

Retroperitoneal mucinous cystic neoplasms, including cystadenoma and cystadenocarcinoma, are rare entities. Few case reports are found in the literature, so the precise incidence is not known[4].

Similarly, to ovarian mucinous tumors, retroperitoneal mucinous tumors are classified histologically into mucinous cystadenomas, borderline mucinous tumors and mucinous carcinomas[5-7].

The first reported case of PRMC was documented by Douglas et al[8] in 1965 and later by Roth et al[9] in 1976.

Different hypotheses have tried to explain the etiopathogenesis of these tumors. One of the hypotheses suggests that PRMCs arise from a retroperitoneal monodermal teratoma. Another hypothesis suggests that these tumors are remnants of the embryonic urogenital tract, in which the cysts develop from specialized mesothelial cells of the urogenital ridge. Other authors postulate a genesis by intestinal duplication. A fourth hypothesis suggests that, since these tumors resemble ovarian mucinous cystic neoplasms, they would arise from ectopic or supernumerary ovarian tissue, although ovarian tissue has not yet been found within a PRMC. Finally, and being the most accepted theory, it is believed that they are produced from invaginations of the peritoneal epithelium during embryonic growth and, subsequently, undergo metaplasia[2,10-12].

The meta-analysis and systematic review by Myriokefalitaki[1] et al in 2015 showed that PRMCs are more frequently observed in premenopausal women, although, as previously mentioned, they have also been reported in men and postmenopausal women. Our patien´s case belongs to this last subgroup. Some authors reported that the mean age of onset is between 42 and 44 years[4,11,13].

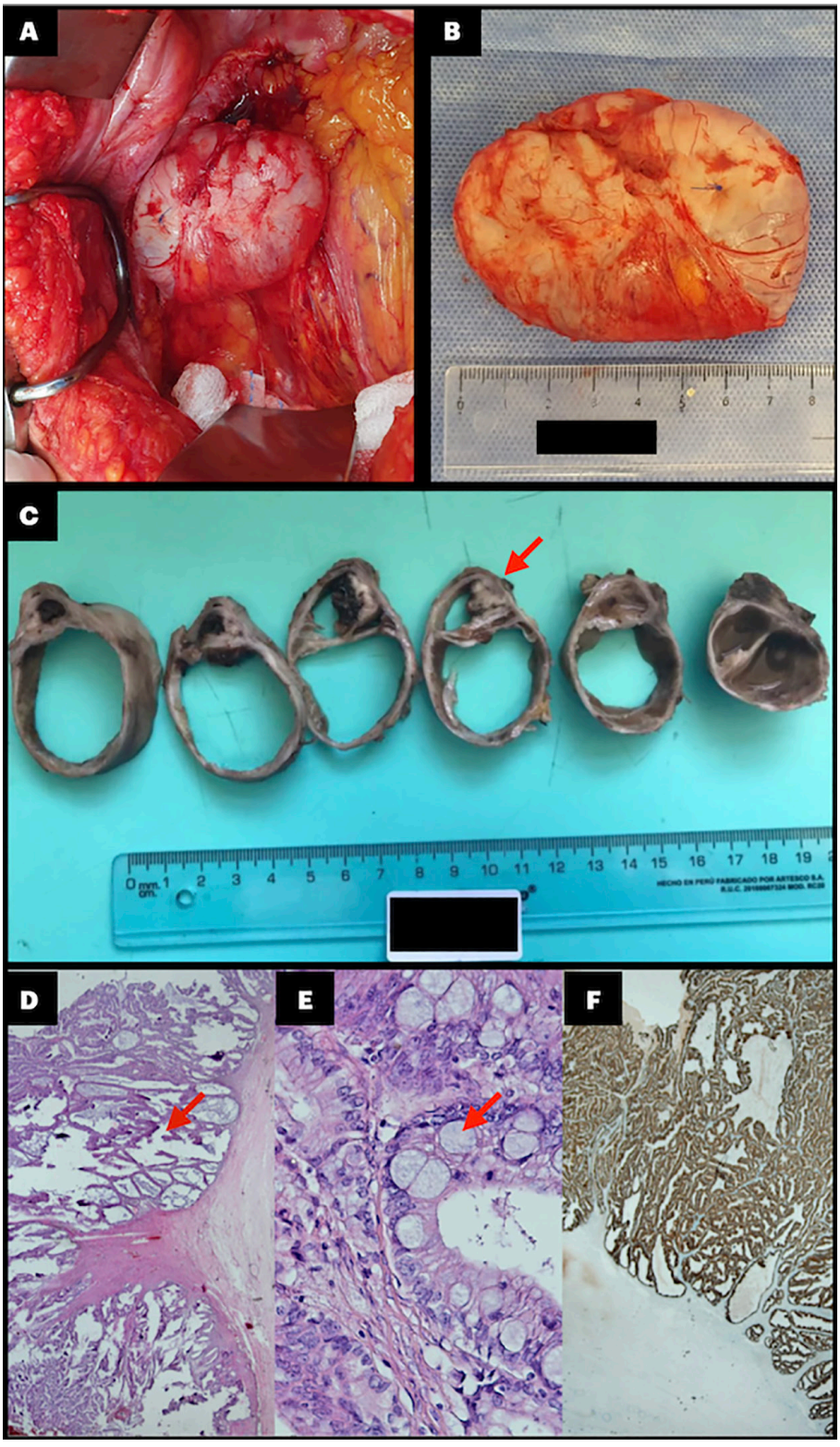
Patients usually present with nonspecific clinical symptoms including abdominal pain, and/or a palpable mass. Our patient did not present any of these symptoms. The diagnosis was reached based on findings on a control abdominal ultrasound due to his underlying nephropathy.

PRMCs present as multilocular or unilocular cystic masses of variable size, located anywhere in the retroperitoneal space[4].

Preoperative diagnosis is challenging due to nonspecific symptoms. Additional imaging studies such as ultrasound, CT scan and MRI are often used to find and locate the tumor. However, these methods cannot differentiate with certainty between a benign and a malignant neoplasm[14], though MRI can identify its mucinous component which would be of diagnostic aid. In our patient, the cyst was identified through an abdominal ultrasound and was characterized by MRI, excluding other differential diagnosis.

Tumor markers, including CEA, CA19.9, CA125, and alpha-fetoprotein (AFP) are non-specific and non-sensitive for diagnosis [1,2,4]. But, in our patient, as in other published cases, they were used to rule out other entities.

Figura 2 | Retroperitoneal mesenteric cyst. A: Exploratory laparotomy, mesenteric cyst in situ. B Fully resected cyst with undamaged walls. C: Macroscopy showing cystic lesion, loculated, smooth internal surface and area with papillary projections (arrow). D: Photomicrograph shows cyst wall with atypical glandular neoplasm with mucin production (arrow) (H&E, 20x). E: Photomicrograph shows glands with goblet cells (intestinal type), with mild pleomorphism (arrow) (H&E, 400x). F: Cytokeratin 7 diffuse positive (immunohistochemical technique 40x)



Differential diagnoses include metastatic mucinous tumors of the ovary, intestine (including the appendix) and pancreas. Benign renal cystic disease, renal lymphangioma or hydatid cysts should also be ruled out[13].

The management of PRMCs is not well established. Surgical excision is the most accepted treatment option[12,15]. For this reason, our patient´s case was discussed in multidisciplinary committee and surgical treatment was chosen, ensuring a complete resection without injuring the cyst.

Adjuvant chemotherapy is not a standard treatment. It is only considered in cases in which the tumor has ruptured during surgery, when invasion to other structures is evident or when metastatic disease is identified[1,10,12].

In the postoperative period, our patient was discussed again in multidisciplinary committee and strict control with images and physical examination every 6 months was decided.

The prognosis of these tumors remains uncertain due to their low frequency and the short patient follow-up. Myriokefalitaki[1] reported a 5-year overall survival of 75.4% for PRMCs. It should be noted that in the majority of reported cases, follow-up did not exceed five years.

More studies will be necessary to determine the correct therapeutic strategy, evaluating long-term follow-up, evolution and prognosis of this disease.

PRMCs are extremely rare tumors, there is limited understanding of its pathogenesis and biological behavior. The diagnosis of PRMCs is challenging due to the absence of specific clinical symptoms, emphasizing the importance of imaging. Management should be decided via expert group after exact review of their preoperative radiological findings. After diagnosis, surgical excision remains as treatment, while the role of adjuvant chemotherapy is limited to specific cases. Periodic monitoring is recommended to detect possible recurrences.

Further studies are needed to enhance our understanding of these rare tumors, allowing for improved management and prognosis.

Conflict of interest: None to declare

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