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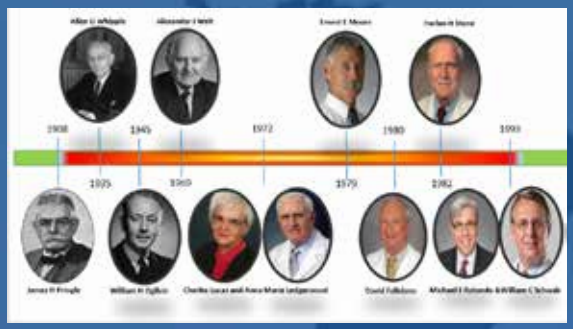

Milestones and Key Figures in the Development of Damage Control Surgery

TRAUMA AND CRITICAL CARE SURGERY UPDATE: EXPANDING THE EVIDENCE — PART II

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Retroperitoneal Mucinous Cystadenocarcinoma

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Abstract

Introduction: Primary retroperitoneal mucinous cystadenocarcinomas (RMC) are rare tumors characterized by mucinous epithelium and ovarian stroma. Their low prevalence complicates the establishment of their etiology, pathogenesis, and behavior. We present a new case of RMC.

Clinical Case: A 32-year-old woman with no relevant medical history presented with abdominal pain and a palpable mass in the right iliac fossa. A CT/MRI scan revealed a 7.5 cm complex cystic mass in the right retroperitoneum, featuring solid formations and in contact with the psoas muscle. Surgery was performed, resulting in the complete resection of the lesion, including partial resection of the psoas muscle and the sciatic nerve. Histological analysis confirmed a poorly differentiated RMC with ovarian stroma, and positivity for PAX8 suggested a Müllerian origin. Five months post-surgery, the patient experienced a recurrence of pelvic pain, and a single iliac lymph node metastasis was identified, prompting the initiation of chemotherapy. Unfortunately, the patient passed away seven months after the surgery.

Conclusion: RMC are rare tumors that necessitate a complex diagnostic approach and multidisciplinary management. Surgery remains the primary treatment, while chemotherapy is considered in selected cases.

Key words: cystadenocarcinoma, mucinous, retroperitoneum, surgery, cancer.

Introduction

Primary retroperitoneal mucinous cystadenocarcinomas (RMCs) are rare cystic tumors composed of mucinous epithelium, typically surrounded by ovarian stroma (1-3). Due to their low prevalence, several hypotheses concerning their etiology and pathogenesis exist, but limited information exists on their behavior (1-3). In this report, we present a new case of RMC.

Clinical Case

A 32-year-old woman with no significant medical or surgical history presented with abdominal pain and a palpable mass in the right iliac fossa. A CT scan revealed a complex multi-septate cystic lesion with solid components measuring 75 x 65 x 66 mm, located posterior to the cecum, ascending colon, terminal ileum, and cecal appendix, and in contact with the iliopsoas muscle (Figure 1). Infiltration of surrounding tissues could not be ruled out. Sub-centimeter lymph nodes were observed in the vicinity of the tumor. An MRI was performed, which showed findings like those described in the CT scan (Figure 2).

In October 2024, she underwent surgery via an infraumbilical median laparotomy. The retroperitoneal lesion was identified during the procedure, confirming that it was not in contact with major blood vessels or the right ureter. The lesion was excised along with a partial resection of the psoas muscle and the sciatic nerve. She was discharged on postoperative day three without complications (Clavien-Dindo 0). As a result of the surgery, she experienced functional impotence and decreased sensitivity in the right leg.

The histological examination revealed a poorly differentiated infiltrating adenocarcinoma within a 7 cm retroperitoneal mucinous cystic neoplasm containing focal ovarian stroma. The lesion tested positive for PAX8, indicating a Müllerian origin. A CT scan conducted in February 2025 due to severe pelvic pain showed lymph node metastasis near the surgical site. As a result, chemotherapy (CBDA-Paclitaxel) was initiated, with plans for early re-evaluation using PET-CT after two cycles.

Figure 1: A CT axial / 1B CT coronal: 7 cm solid-cystic lesion. T: tumor P: psoas

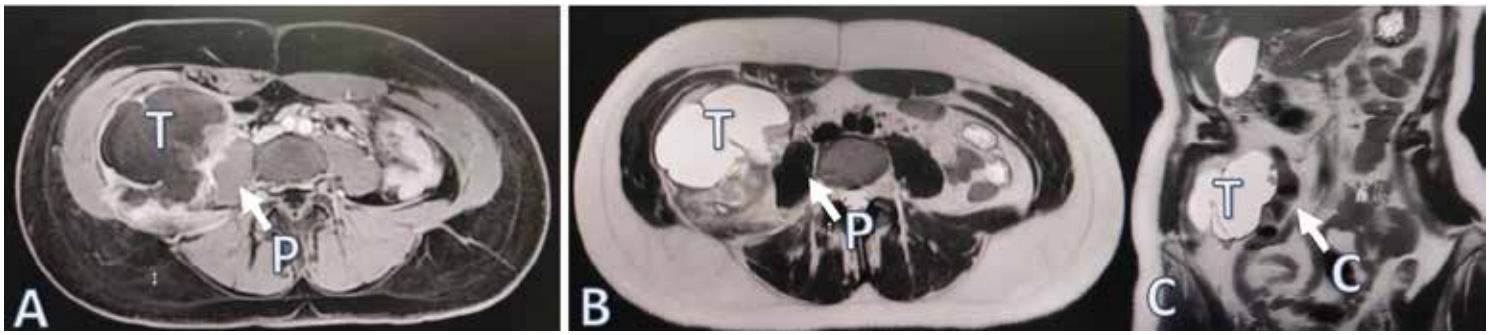
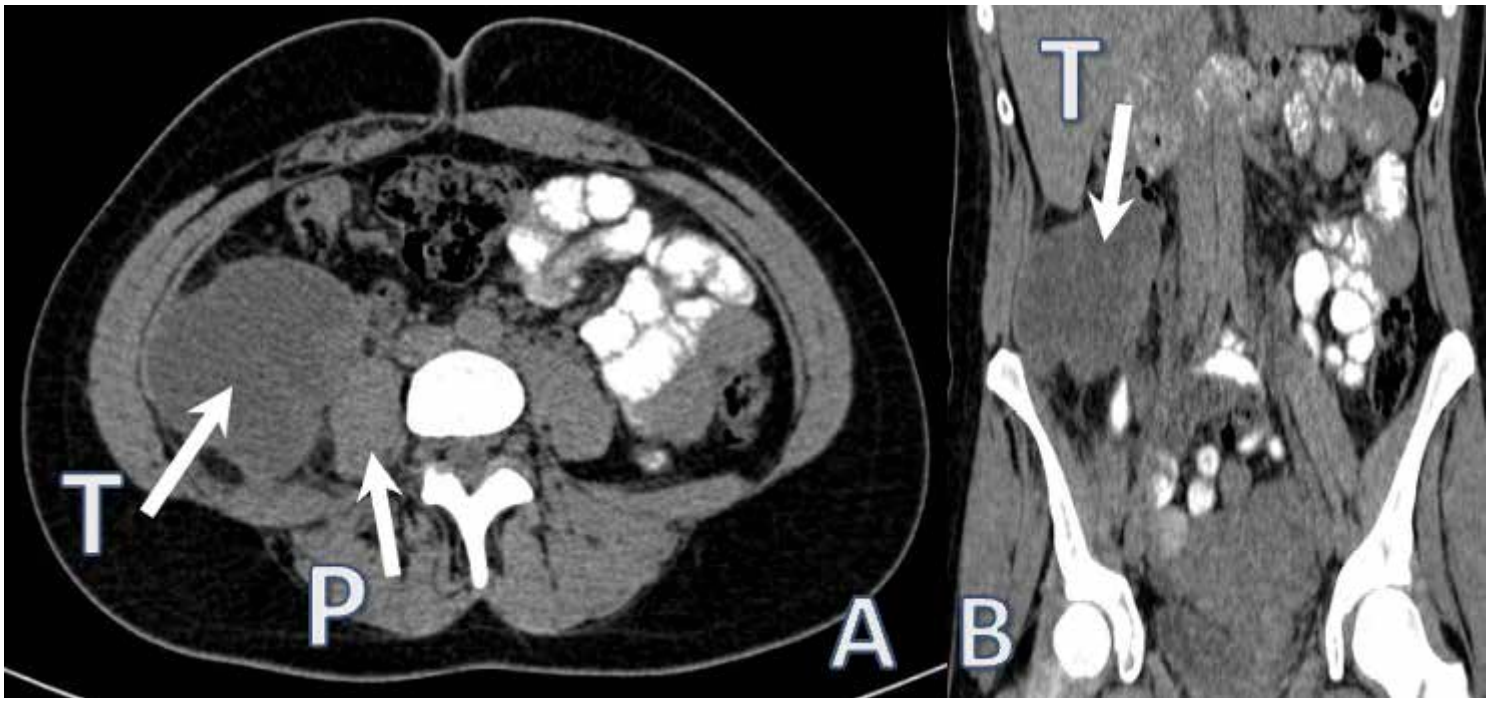


Figure 2: A: MRI axial T1; B: MRI axial T2. C: MRI T2 coronal. 7 cm solid-cystic lesion. T: tumor P: psoas C: Cecum



Unfortunately, disease progression was observed during this assessment. The patient passed away seven months after the surgery.

Discusión

RMCs are a rare type of neoplasm, and their prevalence, pathogenesis, and behavior are poorly understood due to their low occurrence. The first case was reported by Douglas et al. in 1965 (4). This tumor is more common in premenopausal women, typically found in those aged between 42 and 44 years; however, our patient was younger than this average (4). RMC is less common in men and postmenopausal women, and when it does occur in these populations, patients tend to be older, typically between 60 and 80 years. The tumor usually presents unilaterally (1).

Several hypotheses have been proposed to explain the origin of RMCs. These include the idea that they are a type of retroperitoneal mono dermal teratoma, that they develop from embryonic remnants of the urogenital tract—where the cyst arises from specialized mesothelial cells of the urothelial crest—that they originate from an intestinal duplication, or that they stem from ectopic or supernumerary ovarian tissue, given their similarity to ovarian mucinous cystic neoplasms (1-3).

The most widely accepted theory suggests that alterations during embryonic development play a key role. During embryogenesis, the coelomic epithelium eventually forms the peritoneal mesothelium, the germinal epithelium of the ovary, and the Müllerian duct. In patients with CMRs, it is believed that cysts composed of this coelomic epithelium may have persisted in the retroperitoneum. Later in adulthood, these cysts could be influenced by inflammatory and hormonal stimuli, leading to their transformation into RMCs (5). Historically, RMCs are classified histologically into three categories: mucinous cystadenomas (benign), borderline mucinous tumors (with potential for malignancy), and mucinous carcinomas (malignant), as observed in the case we present (1).

The diagnosis of RMCs is challenging and complex. These cysts may be discovered incidentally during imaging tests conducted for unrelated reasons or after a patient notices an abdominal mass through self-examination. Consequently, patients can be asymptomatic or may present with nonspecific symptoms such as abdominal pain, nausea, or a sensation of fullness. The most utilized diagnostic imaging techniques are CT and/or MRIs, which help determine tumor location, size, and the relationship with adjacent structures. However, these imaging methods do not distinguish between benign and malignant neoplasms (1).

RMCs can be either unilobular or multilobular and vary significantly in size, ranging from approximately 3 to 28 cm. They can occur anywhere within the retroperitoneal space (3). Additionally, laboratory tests and tumor markers are generally unhelpful for achieving an accurate preoperative diagnosis, as they lack specificity. In a surgical specimen, next-generation sequencing identified mutations in the KRAS and GNAS genes (3).

The differential diagnosis for this condition includes several possibilities: metastasis from a mucinous tumor of the ovary, intestine, or pancreas; intestinal duplication cyst; cystic kidney disease; renal lymphangioma; and hydatid cysts, among others (3). Preoperative biopsy has limited value, as it rarely leads to a definitive diagnosis, cannot differentiate between benign and malignant tumors, and may even cause tumor dissemination.

The most widely accepted treatment is complete surgical excision, ensuring that the cyst is not ruptured to minimize the risk of spread. Adjuvant chemotherapy is typically not employed unless higher-risk factors exist, such as spontaneous or surgical cyst rupture, invasion of nearby structures, or metastatic disease (5-6).

It is difficult to determine the prognosis for these tumors due to the limited number of published cases and the short follow-up periods (3). Toffolo et al reported a 5-year survival rate of 75.4% when considering all tumor stages (4). However, our patient experienced rapid progression after surgery and had a very short survival rate.

In conclusion, RMCs are rare, and there is still limited understanding of their characteristics. Diagnosing these tumors can be challenging due to their nonspecific clinical features, which underscores the importance of imaging tests. A multidisciplinary team should be responsible for management decisions. Surgical treatment is the primary therapeutic approach, with adjuvant chemotherapy considered in select cases. There is much more to learn about these tumors to enhance diagnostic and treatment strategies.

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