Revista Médica del
Hospital General de México

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CLINICAL CASE

Primary retroperitoneal mucinous cystic retroperitoneal neoplasm with incidentaloma (adenocarcinoma) in the renal capsule

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Abstract

Primary retroperitoneal mucinous neoplasms (PRMCNs) are infrequent and have a remarkable prevalence in women, with a ratio of 9.4:1 compared to men. This statistic is especially relevant today, as this condition was previously believed to be exclusive to women. We present the clinical case of a 60-year-old male patient with a retroperitoneal tumor. A retroperitoneal tumor resection was performed, revealing a mucinous, lobulated tumor surrounding the ureter and displacing the right renal unit and great vessels. As physicians, it is important to take into account this pathology and not to exclude its incidence in men. According to an article published in 2019, 19 cases of PRMCN have been reported, of which two correspond to men. In conclusion, primary retroperitoneal mucinous cystic neoplasms present a challenge for pre-operative management due to the lack of specific data available on this entity.

Keywords: *Retroperitoneal mucinous neoplasms. Retroperitoneal tumor. Primary neoplasm.*

Introduction

Primary retroperitoneal mucinous neoplasms (PRMCNs) are rare tumors, predominantly in the female population. Most neoplasms are found in the left or right lateral retroperitoneal space[1,2]. Histologically, they resemble ovarian mucinous tumors and are similarly classified into the following three categories: benign mucinous cystadenoma, borderline mucinous cystadenoma, and malignant mucinous cystadenoma[3,4]. Pre-operative diagnosis is often confusing due to vague and non-specific clinical signs and symptoms, and the inability of radiology to determine the exact site of origin[5]. The etiology, biology, pathogenesis, and prognosis of PRMCN are still unclear since there is originally no epithelial tissue in the retroperitoneum[3,5]. Several hypotheses have been formulated with the purpose of clarifying the potential origin of these neoplasms. However, to date, some theories have gained greater acceptance within the community than others, underscoring the continued need for rigorous research and thorough analysis to accurately discern the mechanisms underlying the genesis of these neoplastic diseases.

We report the case of a 60-year-old male patient with a history of previous surgeries, who presented with non-specific and intermittent abdominal pain. Following clinical evaluation and imaging studies, a diagnosis of PRMCNM was made, a rare entity that is

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Date of reception: 06-07-2024
Date of acceptance: 22-01-2025
DOI: 10.24875/HGMX.24000045

Available online: 26-08-2025
Rev Med Hosp Gen Mex. 2026;89(1):49-52
www.hospitalgeneral.mx

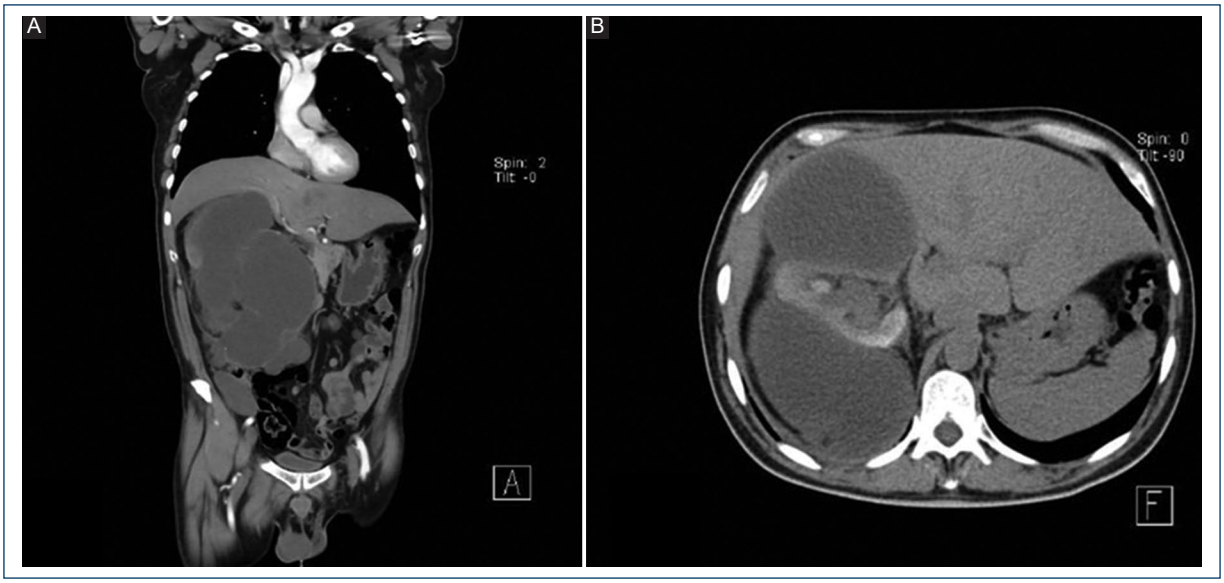

Figure 1. The computed tomography scan reveals a well-defined mass in the right retroperitoneum with a cystic, lobulated, and heterogeneous appearance, likely with mucinous features typical of neoplasms with mass effect. **A:** axial view; **B:** coronal view.

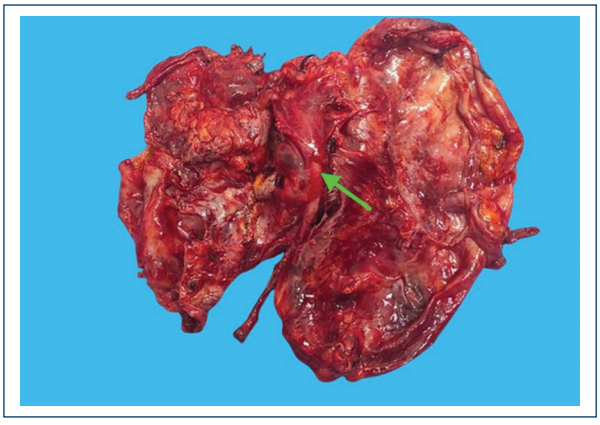

Figure 2. Retroperitoneal tumor capsule. The arrow indicates the ureter attached to the retroperitoneal tumor capsule.

difficult to diagnose due to its vague symptoms and the lack of consensus regarding its origin and pathogenesis.

Clinical case

A 60-year-old male patient with a significant history, open appendectomy secondary to acute appendicitis (2001), left arthroplasty secondary to gonarthrosis (2016); both procedures are reported without complications. The patient began suffering 4 months before surgery, with non-specific and intermittent abdominal pain predominantly in the right hypochondrium, reporting a pain intensity of 4/10 on a Visual Analog Scale, dull type without irradiation or aggravating factors, with increased volume of the right hemiabdomen and data of intestinal pseudo-occlusion, so he went for evaluation. On admission, the patient denied pain, vomiting, fever, hematuria, lithuria, pyuria, or alterations in voiding mechanics. On physical examination, inspection revealed a globular abdomen due to adipose panniculus, with increased volume in the right hemiabdomen without color changes; on auscultation normoactive peristalsis; soft and depressible abdomen, not painful on palpation, reducible, without data of peritoneal irritation, palpable in the right hemiabdomen, non-painful tumor, bilateral Giordano negative. Therefore, a computed tomography (CT) scan was performed, which revealed the right renal unit with contrast medium uptake, failing to visualize the ureter in excretory phase, hypodense retroperitoneal lobulated image of 8.9 × 5.6 × 10.91 cm with altered morphology, displaced toward the ipsilateral diaphragm, with change in situation and anatomy. There is evidence of pyelocaliceal dilatation, and a hypodense, lobulated image in the right ureter topography with dimensions of 19.7 × 8.9 × 20.8 cm with a density of 14.21 HU (Fig. 1). As a complementary diagnostic study, cystoscopy was performed, revealing multiple hypervascularized trabeculations in the bladder that obstructed the

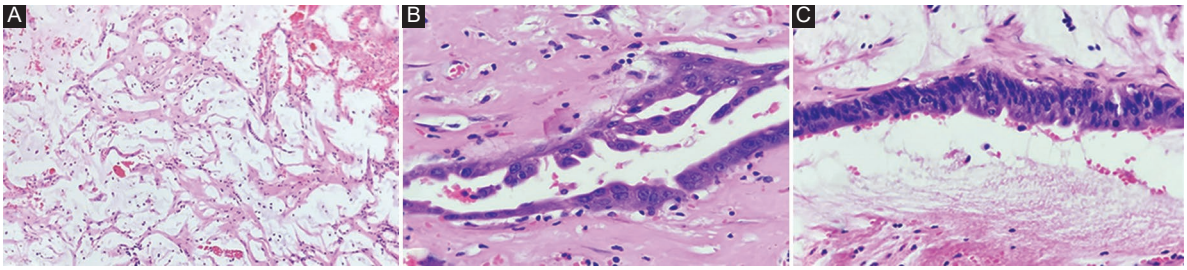

Figure 3. Microphotographs stained with the hematoxylin and eosin method. **A:** fibroconnective tissue with mucin (magnification ×10). **B:** epithelium with high-grade dysplasia (magnification ×40). **C:** dysplastic epithelium (magnification ×40).

visualization of the ureteral meatus. A protocol for surgical treatment of the neoplasm was established in which a simple open right nephrectomy was performed with resection of the tumor with mucinous content (Fig. 2), lobulated, surrounding and adhering to the ureter making its release impossible, displacing the right renal unit and great vessels, approximately 4,000 CC of mucinous material were drained. The patient had an adequate post-operative recovery and was discharged after a 4-day hospital stay, with follow-up care provided at an outpatient clinic. The pathology report describes cystic mucinous neoplasm with high grade dysplasia with focus in a capsule of 5 mm × 4 mm in retroperitoneal space; a peritoneal fluid sample was taken where malignant cells were reported (Fig. 3). Follow-up by the oncology department, where a study protocol was initiated in search of the origin of the primary tumor. Due to the pathological report describing a focus of invasive adenocarcinoma in the capsule of the mucinous neoplasm, a study protocol was initiated, including the evaluation of tumor markers CA 19-9 and carcinoembryonic antigen, both of which were within normal limits. The patient has been under close follow-up by the oncology department for 2 years, undergoing imaging studies (CT scans) every 6 months, along with tumor marker assessments, to detect any potential recurrence or disease progression at an early stage. So far, all results have been negative, and the patient has shown favorable clinical progress with no signs of relapse. Outpatient monitoring will continue as per medical recommendations to ensure long-term disease control.

Discussion

PRMCNs are very rare tumors, are uni or multiloculated cystic tumors with a fibrous capsule and lined by a mucin-producing epithelium, associated with a subepithelial stroma, which was initially described as similar to the ovary[6]. The first case was reported in 1889 by Bassini, where he described a case of cystic adenoma morphologically resembling a mucinous cystadenoma of the ovary[5], and the first male patient with PRMCN was described in 1994. The prevalence of PRMCN among resected retroperitoneal neoplasms was 1.95%[6]. They have a female predominance with a female-to-male ratio of 9.4:1 and are most frequently seen in young adults[5]. The median age at diagnosis was 42.0 years (range 18-86 years), with females being significantly younger than males at diagnosis (42.0 years vs. 62.2 years, p = 0.005)[7]. Despite efforts to understand the pathogenesis of these neoplasms, no definitive conclusion has yet been reached. Several explanatory theories have been proposed, with four of them standing out for their relevance, prominence, and acceptance in the scientific literature. The first explanation is that PRMCs arise from teratomatous lesions, in which columnar epithelium becomes the predominant cell line[8,9]. The second possibility is that they arise from ectopic ovarian tissue[8]; however, ovarian tissue has never been found within a retroperitoneal mucinous cystadenocarcinoma[8,10]. The third hypothesis is that the tumors are remnants of the embryonic urogenital apparatus, in which cysts develop from specialized mesothelial cells of the urogenital ridge. The latest and most widely accepted theory is that PRMCs are produced by invaginations of the peritoneal epithelium during embryonic growth and subsequently undergo metaplasia[8]. An incidentaloma is an unexpected finding. In this case, it was discovered incidentally during the histopathological examination of the renal capsule, revealing a malignant adenocarcinoma localized in this structure. The clinical presentation of this pathology exhibits remarkable specificity and variability, which implies

considerable difficulty in pre-operative diagnosis. The absence of specific laboratory tests, pathognomonic signs, or distinctive radiological findings makes it difficult to discriminate precisely between neoplasms of renal, ovarian (in female patients), or other possible origins. In the different sources consulted, CT of the abdomen is described as the test of choice to evaluate the extension of the tumor, its distribution, and to carry out surgical planning. It has been described that exploratory laparotomy with radical resection without effusion or rupture is the standard therapy and the most important prognostic tool[5,7]. It is definitely important to remove these tumors without rupturing them, and laparoscopic removal should be avoided if the histology of the tumor is unclear[11]. Regarding the application of adjuvant treatment after resection, it is not established due to its limited reported evidence. It is very important to make the diagnosis in time as well as the treatment, since there may be complications such as rupture, infection, and malignant transformation, making the prognosis more unfavorable. The prognosis of patients with these tumors remains uncertain due to their rarity and the fact that most patients were not followed up for 24 months[3].

Myriokefalitaki 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 5 years[7,12].

Conclusion

PRMCN is complex to diagnose due to the lack of specific evidence. There are no radiological patterns or serum markers that initially suggest this entity, and biopsy is risky due to the possibility of tumor dissemination. The challenge lies in determining the type of tumor before surgery, which allows for more accurate and less risky surgical planning. This is crucial to avoid complications such as cyst rupture, which occurred in our patient.

Funding

The authors declare that they have not received funding.

Conflicts of interest

The authors declare no conflicts of interest.

Ethical considerations

Protection of human subjects and animals. The authors declare that no experiments on humans or animals were performed for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from patient, and secured approval from the Ethics Committee. SAGER guidelines have been followed as applicable to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing or creation of the content of this manuscript.

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