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Case report

Surgical resection of a rare primary retroperitoneal mucinous borderline tumor of Müllerian Origin: A case report

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1. Introduction

Primary retroperitoneal mucinous tumors (PRMTs) are a group of rare neoplasms that consist of three subtypes: mucinous cystadenomas, mucinous borderline tumors or tumors of low malignant potential, and mucinous carcinoma. While the pathogenesis remains unclearly defined, PRMTs are believed to arise from mucinous metaplasia of invaginated mesothelium into the retroperitoneum. These rare tumors largely resemble their ovarian counterparts; however, they lack evidence of ovarian tissue upon pathologic review. Establishing a diagnosis of PRMTs preoperatively is challenging, as diagnosis largely occurs in the postoperative setting. Herein, we report the case of a 60-year-old female who presented with abdominal distention, pelvic pain, and bloating with a newly discovered 18 cm retroperitoneal cystic lesion on cross sectional imaging. Pathological findings confirmed a mucinous borderline tumor of Müllerian origin after surgical resection.

2. Case presentation

A 60-year-old Indonesian-speaking female presented for evaluation of a left-sided abdominal mass detected during physical exam by her primary care provider, coincident with a recent history of abdominal distension, non-specific pelvic pain that radiated to her groins bilaterally with post-prandial bloating. She denied changes in her bowel movements, fevers, fatigue or significant weight loss. Past medical history was non-contributory without a prior surgical history. With suspicion of an abdominal mass on physical exam, a CT scan revealed an 11 cm × 14 cm × 18 cm, well-defined ovoid fluid density in her left lower abdomen/retroperitoneum (Fig. 1). A preoperative colonoscopy demonstrated no synchronous disease-related pathology. With concern for underlying malignancy, the patient proceeded to surgical resection.

Exploratory laparotomy revealed a well encapsulated, circumscribed, cystic-appearing lesion emanating from the left retroperitoneum just medial to the left psoas musculature involving but not infiltrating into mesentery of the rectosigmoid. There was no further gross pathology identified in the abdomen or pelvis, including the adnexa, uterus, ovaries, appendix, as well as small and large bowel; all not requiring resection. Surgical resection was limited to the mass, including closely associated peritoneum and mesentery. En bloc resection of bowel was not required, as the lesion appeared in situ, grossly non-infiltrating, and amenable to mesenteric/bowel sparing surgery.

Gross examination revealed a large 18 cm cystic mass with a smooth outer surface. Further inspection revealed a unilocular cyst containing brown watery fluid and cyst wall with mostly smooth inner lining and a focally raised papillary region. Microscopically, the cyst lining showed atypical epithelial proliferation with architectural complexity, including tufting and villous or papillary formation in thickened regions (Fig. 2). The tumor was composed of mucinous gastrointestinal-type epithelium with mild to moderate cytologic atypia, nuclear crowding or stratification, and goblet cells. There was no evidence of invasion into surrounding dense fibrous stroma. No ovarian tissue was identified. Immunohistochemical stains showed tumor cells positive for CK7 (strong, diffuse), PAX8 (patchy), and CK20 (focal), and negative for CDX2 and SATB2. Pathological findings supported a diagnosis of a mucinous borderline tumor of Müllerian origin. Postoperative course was uneventful and the patient discharged on the second postoperative day. Follow-up imaging of the abdomen and pelvis seven months postoperatively remains without evidence of disease recurrence (Fig. 1).

3. Discussion

Primary retroperitoneal mucinous tumors (PRMTs) are a group of rare neoplasms that consist of three subtypes: mucinous cystadenomas, mucinous borderline tumors of low malignant potential, and mucinous

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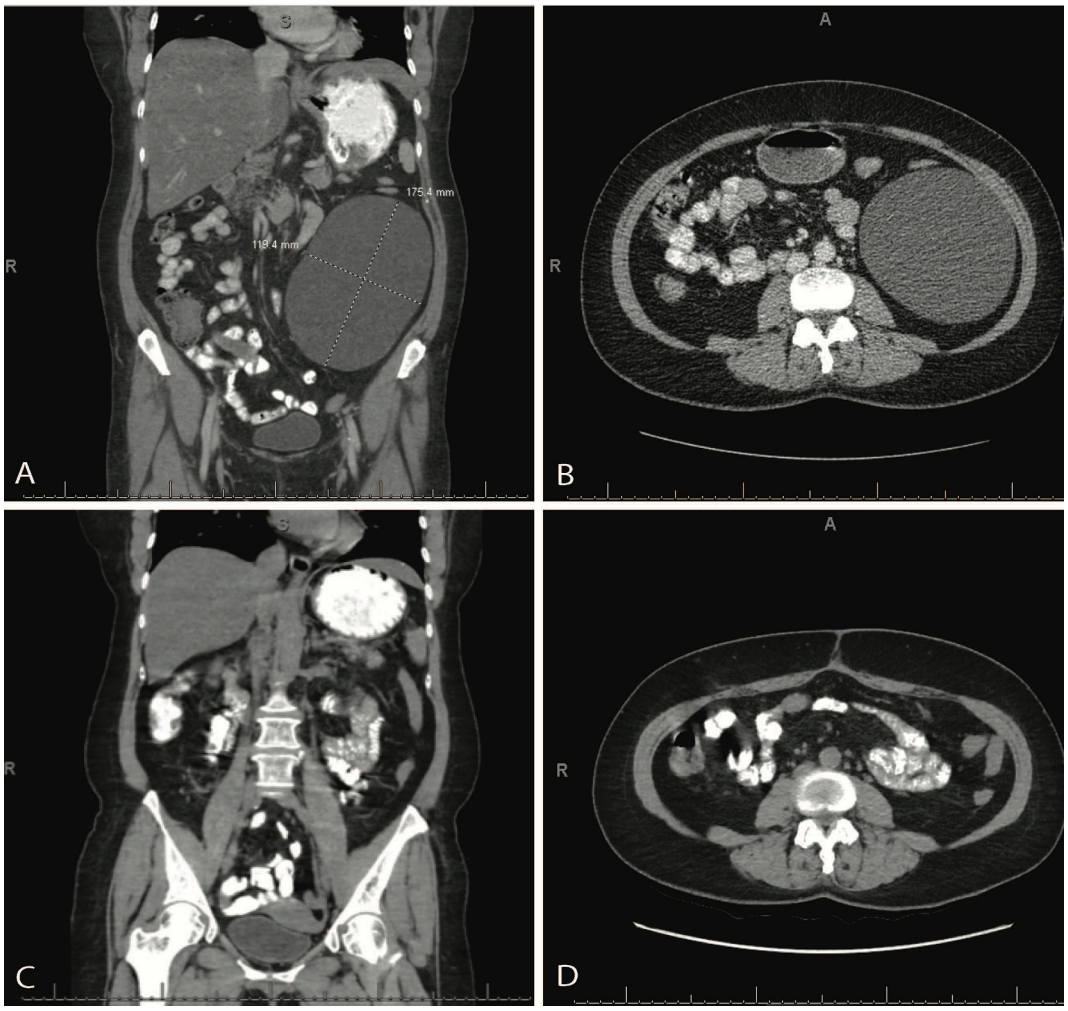

Fig. 1. **A-B** Contrast-enhanced preoperative CT scan of the 18 cm fluid-filled cystic mass in the left abdomen (**A** coronal, **B** axial) concerning for potential malignancy. **C-D** Contrast-enhanced CT scan 7 months postoperatively demonstrating no evidence of recurrent disease (**A** coronal, **B** axial).

carcinoma. To our knowledge, there are approximately 150 cases reported in the world literature, to date. PRMTs are typically asymptomatic with clinical manifestations arising as they enlarge, with the most common symptoms related to mass effect including abdominal distention and pain (Myriokefalitaki et al., 2016). Vastly occurring in women, PRMTs presenting in males are rarely described (Thamboo et al., 2006; Benkirane et al., 2009).

Herein, we discuss the case of a 60-year-old female who presented with several months of abdominal distension, post-prandial bloating, and pelvic pain, later found to have a large cystic mass on imaging and subsequently underwent surgical resection (Fig. 1). Histopathologic evaluation confirmed the presence of a mucinous neoplasm that was morphologically and phenotypically Müllerian in origin, staining positive for CK7 and PAX8 on immunohistochemistry; however, lacking pathologic evidence of true ovarian tissue (Fig. 2). Notably, the patient had grossly normal ovaries and adnexa during the time of surgery. Importantly, features of malignancy including marked cytologic atypia, confluent or solid tumor growth, and stromal invasion were absent.

Histologically, these tumors resemble ovarian mucinous neoplasms; however, lack of true ovarian tissue is requisite for pathologic confirmation (Bifulco et al., 2008; Subramony et al., 2001). Indeed, the origin of these tumors is largely unknown, with three theories of pathogenesis predominating. One theory proposes that these tumors arise from a teratoma with overgrowth of the mucinous component, while another favors a neoplasm originating from heterotopic ovarian tissue (Acharya et al., 2016; Paraskevakou et al., 2014; Roma and Malpica, 2009). This latter etiology holds less favor as most reports note an absence of associated ovarian stroma. The most widely supported etiology favors mucinous metaplasia of the overlying peritoneal mesothelium with resultant cystic mass formation (Subramony et al., 2001; Tenti et al., 1994; Isse et al., 2004). This is based on precedent that peritoneal mesothelium has potential for Müllerian differentiation. Another possibility is that these tumors may originate from remnants of the embryonal urogenital apparatus.

Confirming diagnosis of these tumors preoperatively is challenging with the rarity of presentation and lack of objective radiographic criteria and diagnostic surrogates. Notably, fine needle aspiration of PRMTs and evaluation of aspirated fluid typically provides a paucity of diagnostic information (Cabello Benavente et al., 2017). Consequently, definitive diagnosis requires surgical resection. At our institution, multidisciplinary consensus remains that if an abdominal mass (solid or cystic) is concerning for premalignancy and/or potentially harbors a cancer, and patient performance status and mass appear amenable to complete surgical removal with curative intent, we typically advocate for upfront resection without preoperative biopsy. A preoperative biopsy of a large cystic lesion without solid components is not recommended. This is particularly relevant in our patient with a symptomatic resectable cystic mass. Importantly, this approach obviates potential biopsy-related risks for spillage and/or iatrogenic tumor cell dissemination. Indeed, PRMT is a diagnosis of exclusion, with more frequently encountered epithelial tumors located in the retroperitoneum including ovarian, gastrointestinal, renal cancers and advanced disease metastases. In cases when a

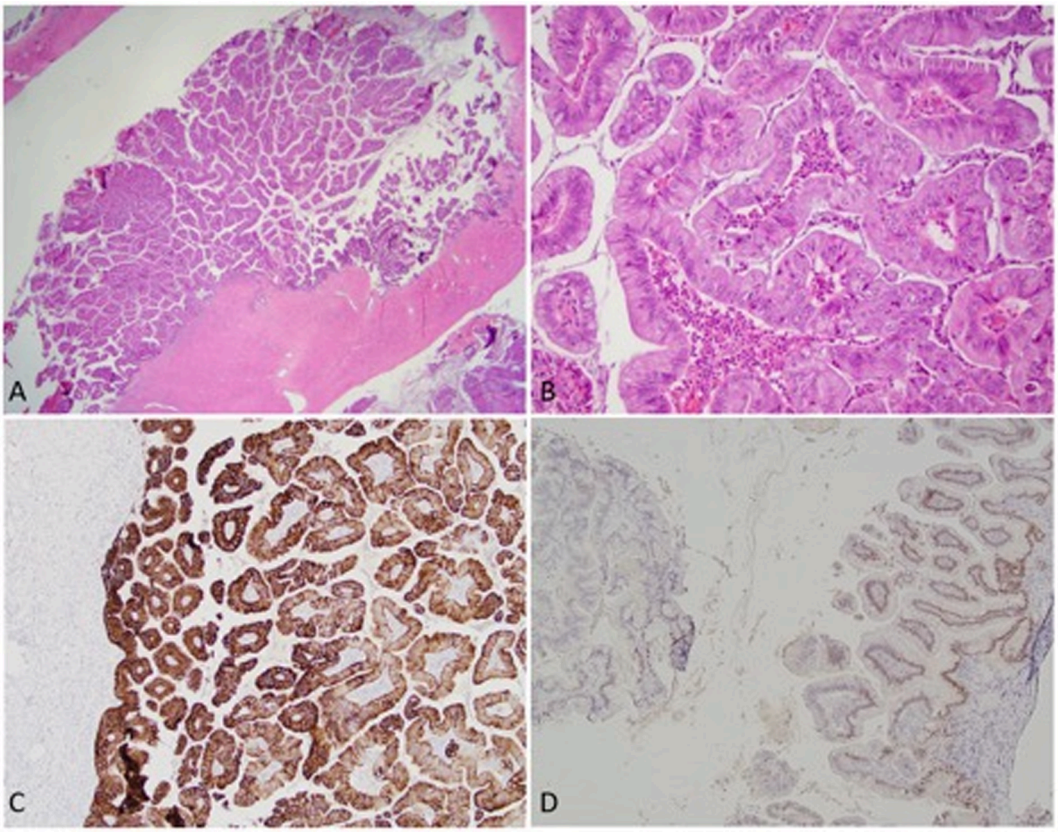

Fig. 2. A. Thickened areas of cyst wall with papillary proliferation (H&E, x20). **B**. Columnar cells with intracellular mucin, occasional goblet cells, nuclear stratification, mild to moderate atypia (H&E, x200). **C-D**. Immunohistochemistry for CK7 (C) and PAX8 (D).

rare, mucin–containing neoplasm is encountered, potential for synchronous colorectal malignancies can occur which necessitates surveillance colonoscopy in appropriate female at-risk patients (Morano et al., 2018).

In general, patients with resectable retroperitoneal masses concerning for malignancy are candidates for surgery with curative intent. Operative treatment includes open and/or minimally invasive strategies that permit careful, safe resection and simultaneous inspection for synchronous disease (Cabello Benavente et al., 2017; Foula et al., 2019). Importantly, cystic fluid has potential to harbor malignant cells, thus surgeons must take extreme care to avoid iatrogenic rupture of the mass during resection. Postoperative treatment considerations are contingent upon pathological review. In our case, this patient with a borderline mucinous tumor underwent surgical resection followed by multidisciplinary recommendation for annual surveillance imaging, thereafter.

Currently, there are no consensus guidelines regarding postsurgical follow-up and surveillance for PRMTs; long-term prognosis for patients with mucinous cystadenomas and mucinous borderline tumors remains favorable, with a 5-year survival reportedly nearing 100% (Wolf et al., 2017). Adjuvant therapy is rarely indicated for borderline tumors (Dayan et al., 2016). In contrast, patients with a diagnosis of mucinous cystadenocarcinoma with reported 5-year survival of 68% necessitates important considerations and strategies for adjuvant therapy (Wolf et al., 2017). Notably, treatment approaches that include hysterectomy, salpingo-oophorectomy, and adjuvant systemic therapy have yet to yield demonstrable survival benefit for this biologically aggressive variant of PRMT.

4. Conclusion

This case report demonstrates a rare patient with a primary retroperitoneal mucinous borderline tumor diagnosed by histopathology after surgical resection. We highlight the importance for consideration of primary retroperitoneal mucinous tumors (PRMTs) in the differential diagnosis when encountering retroperitoneal cystic lesions. Surgical resection provides durable disease control for mucinous borderline tumors or tumors of low malignant potential, as seen in our patient and reported in the existing literature. Limitations of this case report include the retrospective nature of this review, duration of follow-up, and requirement for more conclusive, larger patient outcome studies to validate this treatment strategy.

5. Consent

Written consent was obtained from the patient for publication of this case report.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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