



Primary Retroperitoneal Mucinous Cystadenoma with Borderline Malignancy Concerning in Male Patient: A Case Report and Literature Review

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Abstract

Background: Primary retroperitoneal mucinous cystadenoma of borderline malignancy (PRMC-BM) represents an extremely rare entity neoplasm especially in male patients. The pathogenesis is still controversial. To be the best, the reviewing English literature, only four cases concerning male patients have been reported in the worldwide so far. Therefore, this case may be the fifth case registered in the literature.

Case Presentation: We describe a case of 65-year-old man presented abdominal extension and remittent pain developed. The CT scan and MR images of abdomen revealed a retroperitoneal cystic mass with homogenous enhancing mass measured about 9 by 6 by 3 cm with adjacent to the right adrenal gland. He underwent laparoscopic resection of the tumor with adrenalectomy, and the PRMC-BM pathologically diagnosed. He was discharged the fourth post-operative day with no any complication.

Conclusion: We present an additional case of PRMC-BM concerning male patient. The preoperative diagnosis for PRMC-BM is very difficult. Clinical practice for management with laparotomic or laparoscopic procedure comprises the diagnostic and resection curative approach with quite good prognosis with free from disease after regular follow-up.

Keywords: Retroperitoneal mucinous cystadenoma; Borderline malignancy; Cystadenocarcinoma; Laparoscopy

Introduction

The Primary Retroperitoneal Mucinous Cystadenoma with Borderline Malignancy (PRMC-BM) is an extremely rare tumor affect in male patients [1-4]. The retroperitoneal borderline mucinous cystadenomas belong to benign tumors and the histological spectrum similar to the ovarian tumor is divided into three classifications as benign forms (mucinous cystadenoma), borderline malignancy, and malignant tumor (mucinous cystadenocarcinoma) [4]. Due to regarding the histogenesis, biological behavior, and optimal strategy of management for the tumors are remain speculative. We herein present a case of a PRMC-BM concerning male patient and review the pertinent literature.

Case Presentation

A 56-year-old man visited at our Emergent Department with complaining of a remittent abdominal pain at the right lateral abdomen region during the recent three weeks. He was admitted with vital signs showed blood pressure was 159/107 mmHg, temperature was 36°C, pulse was 96/min, respiratory rate was 22/min. His systemic diseases with medication included anti-hypertensive and gout. By that time, he was under medication with antipsychotic induced Parkinsonism. He has no eventful surgery, no history of alcohol consumption or smoking. He did not fell any evacuation or urination problems. No any allergy history to drugs or food or travelling history mentioned. Physical examination, he revealed alert with chronic ill-looking. Abdomen showed normal bowel sound, no diffused tenderness, no muscle rigidity, no caput medusa, no shifting dullness, no palpable liver or spleen, no Murphy's sign. Genitourinary condition was no cost over tebral angle knocking tenderness. The laboratory findings showed normal within limits and serological tumor

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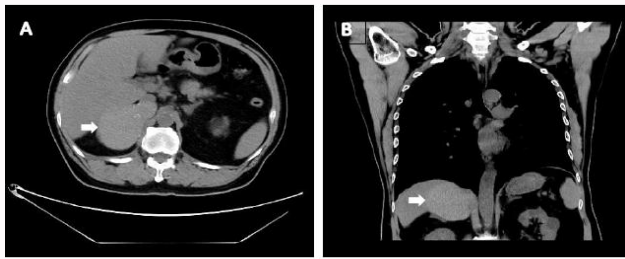


Figure 1: Axial (A) & coronal (B) CT with non-contrast images show right retroperitoneal mass (arrow).

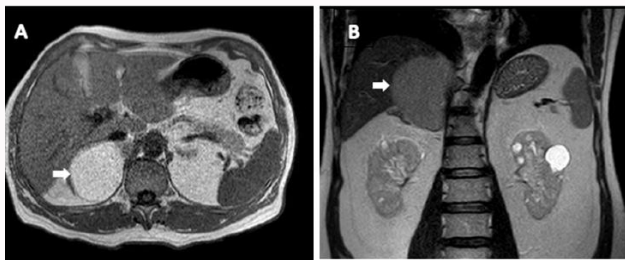


Figure 2: Axial T1WI MR and coronal T2WI MR images demonstrate right suprarenal mass (A, arrow) and lateral displaces the right adrenal gland (B, arrow).

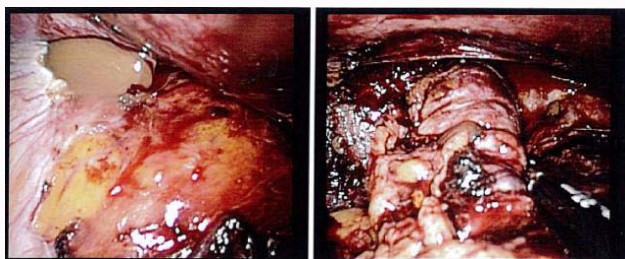


Figure 3: Laparoscopy with complete surgical excision of the right retroperitoneal tumor.

marker including CEA, PSA, AFP, and CA19-9 levels were within the normal reference ranges. The abdominal Computed Tomography (CT) scan images showed a suprarenal cystic mass measured 9 by 6 by 4 cm (Figure 1). The Magnetic Resonance Imaging (MRI) of the abdomen demonstrated suprarenal mass lesion and lateral displaced the right adrenal gland with focal punctuate calcifications (Figure 2). No intratumoral hemorrhage/fluid-fluid level and cystic aspect with some heterogenic focuses also identified. Subsequently, he underwent laparoscopic resection of the tumor (Figure 3). The patient was discharged the fourth post-operative day with no any complication. No evidence of disease recurrence was observed after follow-up 12 months.

Pathological findings

The macroscopic examination of the resection of supra-renal retroperitoneal cystic tumor measured 9 by 6 by 4 cm. The cystic tumor mass showed well-circumscribed and multilobulated characterized by variable thin-walled cysts filled with gelatinous-like or mucin content fluid with surgical opened status (Figure 4A) attached adrenal gland. There also presented focal partly solid, lobulated mass in inner surface of the cystic wall with non-remarkable in grossly, and surrounded by an intact thinned capsule. The adrenal gland displayed unremarkable grossly. Microscopically, the retroperitoneal suprarenal multilobulated cystic mass was characterized by lining

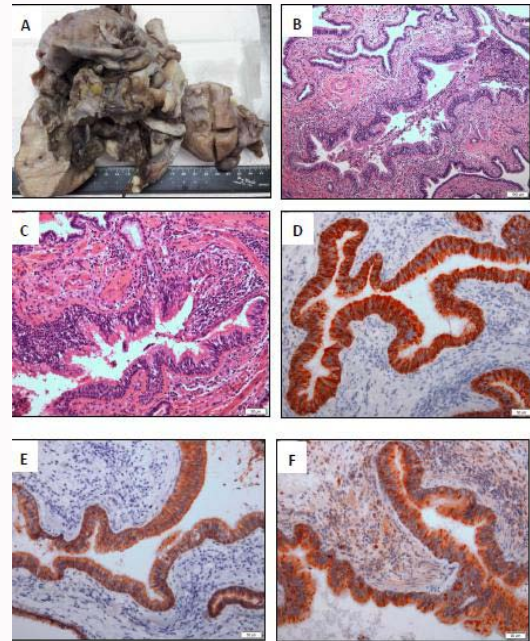


Figure 4: The laparoscopic resection of right supra-renal retroperitoneal multilobular cystic tumor (A). The hematoxylin and eosin staining (H&E) shows a multilobular cystic lesion (B, H&E, original magnification x 10) with an intestinal type mucinous epithelium with focal nuclear stratification and occasional papillary tufts and mild cytological atypia and scant mitotic figures were present (C, H&E, original magnification x 20). The intestinal immunophenotype with immunohistochemical staining demonstrates positive staining for CK7 (D, original magnification x 20), CK20 (E, original magnification x 20) and focal positive for CDX2 (F, original magnification x 20).

with a single layer of both endocervical-like and intestinal-type columnar cells with/without atypia lined the cystic wall (Figure 4B). Focal increased cellular layers, multiloculated architectures greater complexity of the glands and focally papillae, budding, non-ciliated epithelial cells and intracellular mucin. There also presented focal papillary tubular glandular architectures with mild to moderately degrees of proliferative activity lining with cellular stratification with mild cell nuclear atypia (Figure 4C). The Primary Retroperitoneal Mucinous Tumor (PRMT) of low malignant potential (borderline) or primary retroperitoneal mucinous cystadenoma with borderline malignancy (PRMC-BM) was firstly considered. The adrenal gland and surrounded fibrous capsule showed negative for tumor involvement. Immunohistochemical (IHC) study, tumor cells demonstrated positive immunoreactivity for pan-CK, CK7, CK20, and CDX2 (Figure 4D-F), EMA, CEA, MUC-1, but negative for calretinin. Scattered immunostaining for Ki-67 labeling proliferating index showed less than 5% of involved tumor cells. Taken together, based on the morphology, immunophenotype and clinical exclusion of metastasis, the Primary Retroperitoneal Mucinous Cystadenoma with Borderline Malignancy (PRMC-BM) in men was final diagnosed.

Discussion

Primary retroperitoneal mucinous tumors (PRMTs) are even uncommon entity account of 0.01-0.2% of all neoplasms [1-7]. Primary Retroperitoneal Mucinous Cystadenoma (PRMC) is a benign and extremely rare tumor. The Primary Retroperitoneal Mucinous Cystadenoma with Borderline Malignancy (PRMC-BM) is firstly described histologically similar to mucinous cystadenoma found in the ovary. It has the same macroscopic, morphologic and ultrastructural

Table 1: Summary of cases Of Primary Retroperitoneal Mucinous Cystadenoma Of Borderline Malignancy (PRMC-BM) reported in male patients in the literature.

No. Reference [Ref.]	Age (years)	Clinical presentation	Tumor location/size(cm)	Pathologic diagnosis	Follow-up period and outcome
Case 1. Motyama et al. [1]	63	Abdominal pain	Right, peri-nephric / 6 × 4 × 3cm	PRMC-BM	NED
Case 2. Benkirane et al. [2]	44	Right abdominal pain with palpable mass	Right, 4 portions of 5 × 4 × 3 and 4 × 4 × 3 and 3 × 3 × 2 and 2 × 2 × 2cm	PRMC-BM	NED after 12 months follow -up
Case 3. Falidas [3]	37	Right lateral abdominal pain with palpable mass	Retroperitoneal mass / 22 × 14 × 4.5cm	PRMC-BM	Unknown
Case 4. Mattei et al. [4]	32	Right abdominal pain	Retroperitoneal mass / 7 × 6 × 4.4cm	PRMC-BM	Unknown
Present Case	65	Right abdominal pain	Right suprarenal mass / 9 × 6 × 3 cm	PRMC-BM	NED after 12 months follow-up

PRMC-BM: Primary Retroperitoneal Mucinous Cystadenoma of Borderline Malignancy; NED: No Evidence Of Disease; Ref. Reference, No.: number.

characteristics compare to the ovary mucinous processes [2,8,9]. PRMTs are divided into three categories similar to the counterparts arising from ovarian mucinous neoplasms as Primary Retroperitoneal Mucinous Cystadenomas (PRMCs), Mucinous Cystadenomas of Borderline Malignancy (PRMC-BM) and Cystadenocarcinomas (PRMC-Cas). PRMC-BM concerning in men is extremely rare tumors, almost exclusively female patients. PRMC-BM belongs to the histological spectrum transforming from benign forms (mucinous cystadenoma) to malignant change (mucinous cystadenocarcinoma) [8-10]. Preoperative diagnosis of the PRMC-BM is very difficult due to the majority of them are malignant characteristics with presenting a non-specific symptomatology and the vague preoperative imaging findings. Until today, the pathogenesis of the development of PRMC-BM is still unknown. The differential diagnosis of the PRMC-BM could be included ectopic ovarian tissue or supernumerary ovary (in women), mucinous cystadenoma, cystic lymphangioma, cystic teratoma origin, lymphocele, cystic mesothelioma, embryonal urogenital remnants, urinoma, intestinal duplication and metaplastic invaginations of peritoneum [1-7].

In retrospective study and reviewing the literature, Lai et al. [11] reported the first case of a 52-year-old male with pure PRMC-Ca. Motoyama et al. [1] reported the first case of a 63-year-old male with PRMC-BM diagnosed by fine needle aspiration cytological procedure. Subsequently, Benkirane et al. [2] presented the second case of a 44-year-old male patient with PRMC-BM, respectively. The third case, Falidas et al. [3] illustrated a 37-year-old man with PRMC-BM. Authors have asserted that the overgrowing of the mucinous epithelium on teratoma or genitourinary remnants constitutes two the peritoneal epithelium possesses the potential of Müllerian differentiation [3,12]. However, recent investigators consent that PRMCs originate from multipotential mesothelial cells entrapped in the retroperitoneum during the differentiation process. These epithelia undergo a mucinous metaplasia and turn into cystic mucinous configuration with cytological changes or malignant transforming phenotypes [3,13,14]. Mattei et al. [4] identified the forth case of a 32-year-old male patient presented a right testicular mass further received the abdominal CT scan and incidentally found a retroperitoneal interaortocaval solid/cystic mass. Subsequently, he underwent a laparoscopic excision completely of the retroperitoneal mass, and the final pathologic diagnosis was PRMC-BM made. Up to date, the reviewing of English literature, our this additional case could be the fifth registered case concerning male patient with PRMC-BM in the worldwide shown as (Table 1). To differentiate diagnosis for the PRMC-BM of the tumor correlate to soft tissue and radiological evidence of its origin, the diagnostic value of Computed Tomography (CT) and Magnetic Resonance (MR) imaging scan is important and preferred diagnostic value in PRMC-BM. The pathogenesis of PRMC-BM is still unclear. PRMC-BM is firstly

described histologically similar to mucinous cystadenoma found in the ovary. PRMC-BM of epithelial origin are rare because epithelial cells are usually not observed in the normally peritoneum and their origin has been controversial. The latest hypothesis relates to the metaplastic origin of the cysts: they may develop from coelomic epithelium [2,13]. During embryogenesis, coelomic epithelium is transformed into the peritoneal mesothelial and ovarian germinal epithelium. Peritoneal mesothelial cells display to maintain the same differentiation characteristics as ovarian epithelial tumors [10]. The most theories about the pathogenesis of PRMC-BM development suggest that invaginations of the peritoneum (mesothelium) owing to inclusion cysts progressing mucinous metaplasia and causing in a mucinous cystadenoma. These results might develop on to borderline and malignant mucinous tumors. Other popular hypotheses include an origin from ectopic ovarian tissue and an ovarian teratoma, where the mucous epithelium has surpassed all other components to become the only identifiable component, or from intestinal replication, also known as enterogenous genesis development [1-4,13].

Previously, some documented reports study by the IHC profiles of PRMCs-BM. Reviewing the literature, Motoyama et al. [1] described that two cases with benign and borderline mucous epithelium demonstrated apical membrane staining for Carcino Embryonic Antigen (CEA), and mucinous cystadenocarcinoma components showed more extensive cytoplasmic immunostaining and concluded the CEA marker pattern is similar to ovarian mucinous tumors. The IHC analysis revealed a positive match to CK7 and CK20 antibodies. This is the same profile assay as in ovarian mucinous tumors [9,11,22]. Thamboo et al. [23] illustrated strongly patchy staining for CK7, CK20, CEA, and CA 19-9 were shown in the benign and atypical epithelium. Previously, CK7, CK20 and CDX2 displayed positively stained suggested the presence of ovarian tissue. The IHC profiles study demonstrated epithelial cells positive for CK 8/18, CK20, pan-CK, CEA and Ki-67 [3,11]. Some investigators believed that female PRMC is derived from the mucinous metaplasia of the peritoneal (mesothelial) inclusion cysts, rather than from ectopic ovarian tissue or ovarian teratoma, due to this tumor do not occur in male patients. In our case, the positive immunostaining for CDX2 and negative for calretinin, so the histogenesis of PRMC is still controversial [1-4]. The similarity between immunohistochemistry and ultrastructure, the characteristics of ovarian mucinous tumors sustain this hypothesis. The first hypothesis suggests that PRMC-BM are derived from a retroperitoneal monodermal teratoma with predominant columnar epithelium [14,15]. Other authors hypothesized that enterogenous genesis occurs due to intestinal replication [15-17]. In our case, intestinal-like epithelial tumors and smooth muscle fibers around the cystic tumor can support this hypothesis. Owing to these tumors like to ovarian mucinous cystic tumors, a third statement is postulated they are derived from ectopic ovarian tissue. However, no ovarian tissue

has ever been found within a PRMC-BM [18]. The fourth possibility is that the tumors are remnants of the embryonal urogenital apparatus, where the cyst is developed by specialized mesothelial cells of the genitourinary ridge [19]. The most generally recognized to be accepted theory is that PRMC-BMs occur in the invagination of the peritoneal epithelium during embryonic growth and subsequent metaplasia [20-22]. Previously, Pennell et al. [24] reported that ovarian tissue was only observed in one reported female case of PRMC, and that ovarian tissue should not be presented in this male case. Additionally, PRMC-BM has also previously reported in male patient. These findings could be ruled out the origin of ectopic ovarian tissue as PRMC-BM. Because of the lack of well-developed intestinal mucosa and smooth muscle in PRMC-BM, enterogenic-induced intestinal replication can also be excluded. Due to the lack of other tissue types or typical structures of teratomas in PRMC-BM, monocortical variants of teratomas are unlikely to occur in the present case. Although the morphological and immunohistochemical features of ovarian mucinous neoplasms and primary retroperitoneal mucinous tumors are similar, the two theories of ectopic ovarian tissue origin and ovarian teratomas origin cannot be characterized clearly the tumors found in male patients, such as under these circumstances. Suggestive of possible source of PRMC-BM concerning in male patient may be an unreduced testicles and can be excluded in our case. The differential diagnoses of patient with PRMC include metastatic mucinous tumors from other sites such as the ovaries (in women), the gastrointestinal tract (including the appendix), adrenal gland, kidney and the pancreas etc. and should be excluded by careful history-taking, clinical examination and diagnostic imaging [1-7]. The exploratory laparotomy or laparoscopy with complete resection of the cystic tumor is preferred for both the diagnostic assessment and management of PRMCs-BM. Owing to the extremely rarely PRMC-BM cases reported in the English literature, several investigators could not suggested or recommend further any adjuvant chemotherapy protocols. The prognosis and survival for majority cases of PRMC is to be good. In previous additional case reports demonstrate that free of disease, no recurrence or metastases on short-term follow-up to 3 months or even for 3 years of life. Because the experience of male with PRMC remains uncommon tumors and their biological behavior and survival will be further evaluated for much more cases are encountered.

Conclusion

We describe an additional extremely rare case of PRMC-BM in a male patient and no evidence of tumor recurrence was detected during a follow-up period of 12 months. Previous investigators and several hypotheses have been proposed the histogenesis for PRMC-BM. The treatment of this tumor requires surgical resections completely is advised for diagnostic assessment and management with a long term follow up, especially borderline foci exists, to prevent relapses or malignant potential developed. To our knowledge and review the literature, the small number of reported cases elucidated and insufficient well-documented surveillance data, the evaluation of prognosis of recurrence is still controversy.

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