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Case Report

Primary retroperitoneal mucinous cystadenoma in a male patient: a case report

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ABSTRACT

Primary retroperitoneal mucinous cystic neoplasms are very rare, especially in men. To our knowledge, only 13 cases of such neoplasms have been reported in men to date. The most common type is the primary retroperitoneal mucinous cystadenoma, which almost invariably affects females. Most patients present with non-specific symptoms. In this case report, we will describe a case of a 53 year old male patient who presented with raised CEA tumour marker and vague right sided abdominal discomfort. A computed tomography (CT) scan was performed, which revealed a 4.9 × 4.7 × 7.5 cm lobulated retroperitoneal cystic lesion abutting the posterior wall of the mid ascending colon. A laparoscopic excision was eventually performed with complete removal of the retroperitoneal cystic lesion. Subsequent histological assessment confirmed the diagnosis of a primary retroperitoneal mucinous cystadenoma with borderline malignancy. On comparison with a prior CT scan performed about seven and a half years earlier, we managed to derive a doubling time of about 626 days, which is the first reported case in English literature to document the growth rate of such a tumour. The patient has since been discharged well, with no evidence of tumour recurrence on an interval CT scan.

Keywords: Primary retroperitoneal mucinous cystadenoma, Primary retroperitoneal mucinous cystic neoplasms, Male, Growth rate, Doubling time

INTRODUCTION

Since its first description in 1924 by Handfield, primary retroperitoneal mucinous cystic neoplasms continue to be extremely rare, especially in males.[1] These neoplasms can be classified into three broad categories: (a) primary retroperitoneal mucinous cystadenomas (most common), (b) primary retroperitoneal mucinous cystadenomas with borderline malignancy and (c) primary retroperitoneal mucinous cystadenocarcinomas. These neoplasms almost exclusively affect females only, with only 13 cases of such neoplasms having been reported in men to date (based on PubMed online database search as on 1st March 2019). Most patients present with non-specific symptoms. Based on reported cases in literature, the most common clinical presentations were either a palpable abdominal mass or abdominal pain/discomfort.[2-4] In this case report, we will be describing a rare case of male primary retroperitoneal mucinous cystadenoma with borderline malignancy and a review of its management.

CASE REPORT

Mr LWT was a 53 years old male with a past medical history of psoriatic arthropathy affecting the small joints of his hands, hypertension and hyperlipidaemia. He presented to the outpatient clinic with a 1 month history of vague right sided abdominal discomfort and raised CEA. CEA test was performed as part of his health screening examination which was raised at 8 µg/l. Physical examination during this current presentation was unremarkable with no palpable mass.

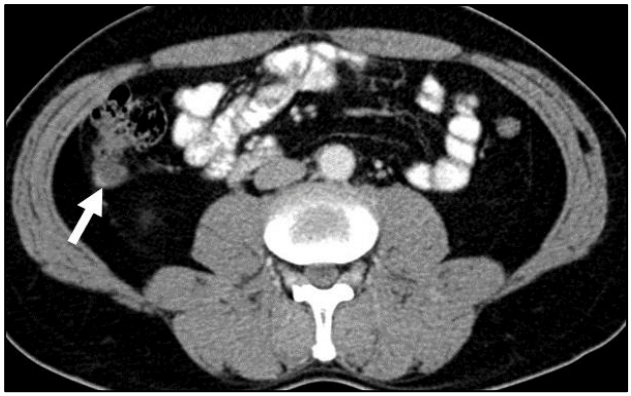

Figure 1: Axial contrast enhanced CT of the mid-abdomen showing a small cystic nodule with hyper dense calcific rim (arrow) that abuts the posterior wall of the ascending colon. The nodule measures 30 cc in volume.

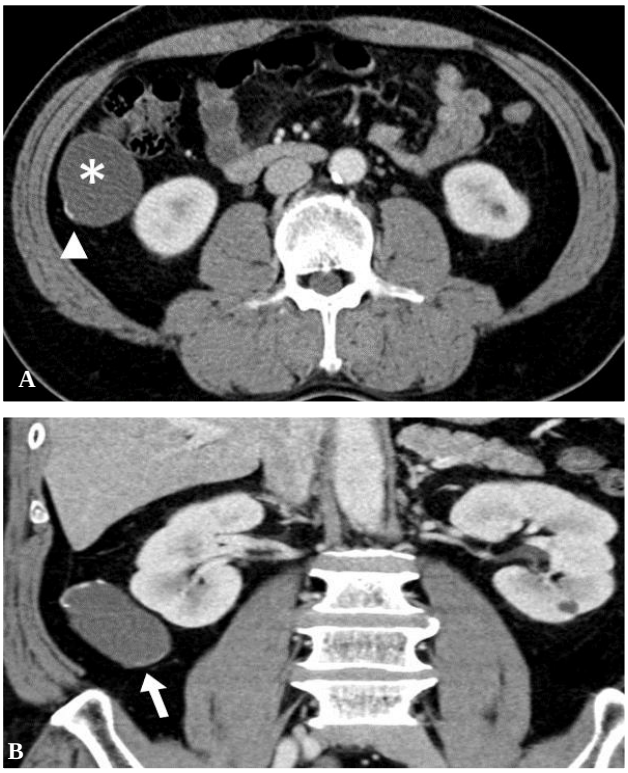

Figure 2: (A) Axial CECT of the same patient eight years later shows the cystic nodule (*) to have increased significantly in size. Rim calcification remains present (arrowhead); (B) Coronal reformatted CECT of the nodule (arrow) shows the craniocaudal extent of the lesion. It is separate from the kidney and there are no infiltrative features into the surrounding retroperitoneal fat. The lesion measures 623.3cc.

Mr LWT also gave a history of a prior computed topography (CT) scan of his abdomen performed at another hospital seven and a half years ago, as part of the investigative work up for his symptoms of abdominal pain then. The CT scan showed a 1.7 × 1.2 × 3.4 cm (30cc) cystic focus abutting and posterior to the ascending colon. No further investigations or treatment was administered after the scan (Figure 1).

A contrast-enhanced CT (CECT) scan of the abdomen and pelvis was performed, which revealed a lobulated 4.9 × 4.7 × 7.5 cm (623.3 cc) lobulated cystic lesions arising within the left retroperitoneal region abutting the posterior wall of the mid ascending colon. There were tiny punctate calcifications noted at parts of the cyst wall. No enlarged abdominal/pelvic lymph nodes or ascites were detected (Figure 2).

An MRI abdomen was also performed to better characterize the lesion, which showed a septated cystic mass measuring about 8 cm in size. No internal mural nodularity or enhancement was seen. The calcifications noted on the CT scan were not apparent on the MRI scan. Overall, the cystic lesion exhibited a benign morphology (Figure 3).

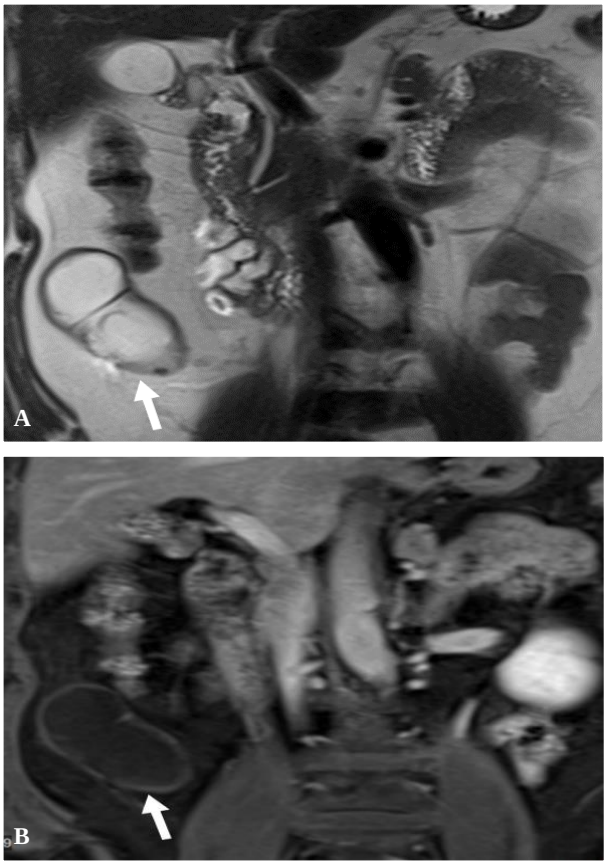

Figure 3: (A) Coronal single shot fast spin echo T2-weighted image of the same lesion (arrow) shows fluid signal within the cyst, and slightly thickened wall with thin internal septation; (B) Coronal T1-weighted fat suppressed gradient recalled echo image of the lesion shows faint enhancement of the wall (arrow). No mural nodules or enhancement of the internal contents. MRI is not sensitive for depiction of the rim calcification seen on corresponding CT.

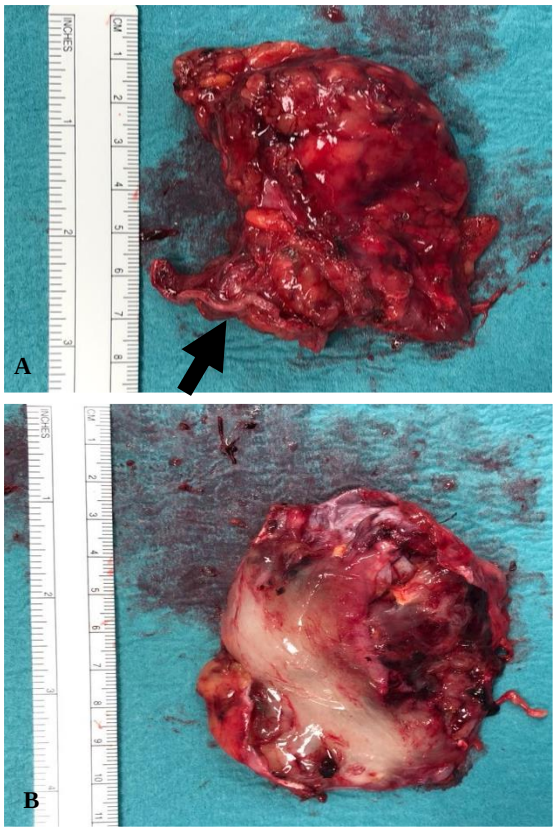

Figure 4: (A) Photo of the resected specimen, with cuff of ascending colon excised en-bloc (black arrow); (B) Photo of the reverse side of specimen. The specimen was soft and cystic in nature with mucous contents discernible through the thin cystic wall.

A contrast enhanced CT scan of the thorax was performed which did not show any other lesions or evidence of metastasis in the thorax. Colonoscopy did not reveal any significant abnormalities. A diagnosis of possible cystic lymphangioma is made.

Mr LWT was offered surgical resection of the lesion in view of the increasing size and to rule out any malignant pathology. He underwent a laparoscopic excision of the retroperitoneal cystic lesion. Intra-operatively, the tumour was densely adhered to the posterior wall of the ascending colon, with no discernible plane. As such, the lesion was excised enbloc with wall of ascending colon to ensure complete removal of the cyst (Figure 4).

Post operatively, Mr LWT made an uneventful recovery and was discharged well on post-operative day 2.

Detailed histological assessment of the cystic specimen showed a cyst wall composing of mucinous epithelial cells overlying hyalinized fibrous issue. The lining epithelium exhibit low grade cytological atypia and apical mucin. In a focal area, the epithelial cells were seen to invade the hyalinized muscularis propria with broad front. Fragments of mucinous epithelial cells were also found in the pools of extracellular mucin in the cyst contents. The histological grade and type were deemed as a well differentiated low grade mucinous neoplasm. No evidence of lymphovascular invasion was seen. The histology confirmed the diagnosis of a Primary Retroperitoneal Mucinous Cystadenoma with borderline malignancy (Figure 5).

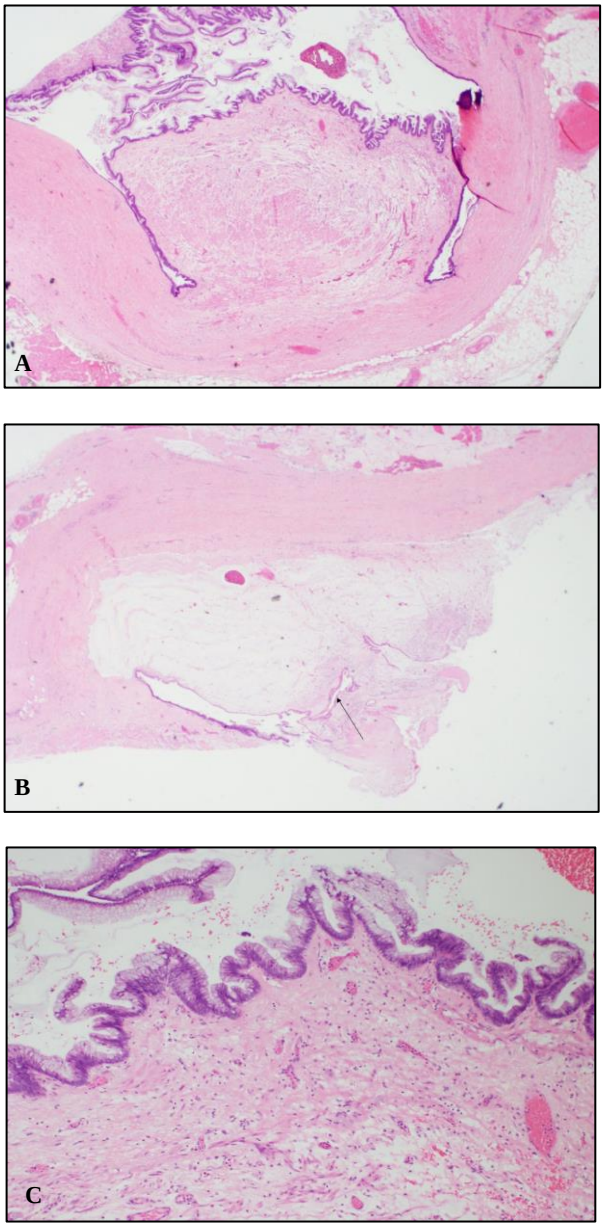

Figure 5: (A) The low power field of the cyst shows pools of extracellular mucin containing fragments of glandular epithelial cells. (Hematoxylin and eosin stain, x10), (B) In a focal area, the epithelial cells invade the hyalinized cyst wall with a broad front (the invasive front is indicated by the arrow). (Hematoxylin and eosin stain, x10), (C) The medium power field shows the mild to moderate cytologic atypia of the epithelial cells lining the cyst (Hematoxylin and eosin stain, x100).

Mr LWT was followed up outpatient and did not exhibit any new abdominal or gastrointestinal signs and symptoms. CEA levels taken at 2 months post-operatively dropped back to within normal range (3 µg/l). A repeat contrast-enhanced CT scan performed 4 months post-operatively did not reveal any evidence of tumour recurrence.

DISCUSSION

Primary retroperitoneal mucinous cystic neoplasms continue to be a very rare phenomenon almost always affecting females only, with very few cases being reported in males.

Since the retroperitoneum does not contain any mucinous epithelial cells, the histiogenesis and development of these tumours still remain unclear. There are several theories explaining the pathogenesis of primary retroperitoneal mucinous cystic neoplasms, including heterotopic ovaries (in females), mucinous metaplasia of the mesothelial lining, teratoma formation, or ectopic/undescended testes (in males).[2-5]

There are currently no pathognomonic clinical or radiological findings for these tumours. Hence, obtaining an accurate pre-operative diagnosis based solely on imaging modalities remains challenging. These tumours can present as uni- or multi- loculated cystic lesions anywhere in the retroperitoneal space - often displacing retroperitoneal and intra-abdominal organs due to their large size. The diagnostic value of MRI and CT is similar; MRI provides a variety of images and can identify correlations between the mass and the soft tissues, whereas CT may shows mural calcifications, which support the diagnosis of cystadenoma rather than teratoma.

Depending on the location, differential diagnoses include cystic teratoma, lymphocele, lymphangioma, cystic mesothelioma, Müllerian cyst, epidermoid cyst, pancreatic pseudocyst, and urinoma. Aspiration and cytology examination generally do not provide much diagnostic benefit as they cannot reveal the type of cells lining these cystic tumours, and carries risk of tumor cells seeding

Very few reports have studied the immunohistochemical profile of these tumours.[6,7] Subramony et al found that stromal cells in PRM cystademonas express the estrogen receptor, which may be a reason as to why these tumours are more frequently found in women; and postulated that the growth of the tumour could be due to activation of the estrogen receptor.[6] Motoyama et al found that the epithelium of benign and borderline mucinous cystadenomas showed apical membranous staining for CEA, whereas the epithelium of mucinous cystadeno-carcinoma showed more extensive cytoplasmic staining with this marker.[8] Due to the rarity of such cases, the expression and significance of CEA remains unclear. In our case, CEA levels were noted to be raised pre-operatively, which subsequently fell back to within normal range after complete excision of the tumour.

Interestingly, our case report is the first case in English literature to have an earlier scan documenting the tumour in its early stages. The CT scan done approximately seven and half years prior shows a tumour volume of 30 cc, which eventually grew to a size of 623.3 cc in the pre-operative CT scan. Assuming that the growth rate of the tumour remained constant, this will give the tumour a doubling-time of 626 days.

Surgery and excision of the tumour continues to be the treatment of choice and only definitive way to confirm the diagnosis in most reported cases, as per our case.[2-4] Complete surgical resection of the tumour is generally advised to prevent recurrence, compressive complications due to mass effect and risk of malignant transformation.

CONCLUSION

This case report is the 14th case of a primary retroperitoneal mucinous neoplasms being reported in a male patient. Similar to previous case reports, our patient presented with non-specific symptoms of abdominal discomfort. Of note, our patient also presented with raised CEA levels, which dropped back to within normal range after removal of the tumour. However, due to the rarity of these tumours, the expression and significance of CEA remains unclear and requires further studying. Due to the lack of pathognomonic radiological and clinical findings, clinching a definitive pre-operative diagnosis of primary retroperitoneal mucinous neoplasms continues to be challenging. Assuming that the tumour's growth rate remained constant, the doubling-time of this particular primary retroperitoneal mucinous cystadenoma with borderline malignancy was 626 days, which can be considered as a slow growing tumour. Being the first and only case in English literature to report the growth rate of such a tumour, the accuracy and clinical significance of this value is yet to be determined. Regardless, it provides us with a better understanding into the inherent growth characteristics and tumour biology of these neoplasms and may help guide in the frequency of post-operative imaging surveillance.

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