

Case Report

Primary Retroperitoneal Mucinous Cystadenoma: A Case Report and Review of Literature

Manisha E Kotay*

American University of the Caribbean School of Medicine (AUC), Sint Maarten, Dunedin, Florida, USA

Abstract

This case report describes an unusual injury in a 15-year-old female who sustained a glenohumeral fracture dislocation with osteochondral shear injury of the Primary Retroperitoneal Mucinous Cystadenoma (PRMC) is a very rare diagnosis. A case of rare PRMC unaffiliated with the ovaries in a reproductive aged woman is reported. The patient had undergone percutaneous drainage five years before, but the mass recurred. The retroperitoneal mass was removed and confirmed by histology to be a benign PRMC. No other intervention was indicated. A diagnosis of PRMC is extremely rare, but very important to diagnose for malignant change is possible. This case shows the typical cantillation of symptoms and a path for recovery. The literature review sheds light on possible etymologies of this rare diagnosis.

Keywords: PRMC; Organs; Tumors; Laparoscopic surgery; Neoplasms

Introduction

Primary retroperitoneal tumors account for less than 0.02% of neoplasms and Primary Retroperitoneal Mucinous Cystadenomas (PRMC) are just one small subset of these tumors [1]. To our knowledge there are around 30 cases reported in the literature [2]. Mucinous Cystadenomas are much more likely to arise in conjunction with ovaries and sometimes pancreas masses. General findings are vague symptoms of a retroperitoneal mass -feelings of pressure and pain in the abdomen sometimes accompanied by obstructing effects on nearby organs [3]. There are so few reported cases that the origin of these masses is still uncertain. Two main theories seem to dominate the discussion. Both of these theories tie retroperitoneal and ovarian mucinous cystadenomas. This originates from the fact that grossly and histologically the two are indistinguishable [4-6]. Both tumors are cystic with mucous secretion inside a thinly lined wall of a single layer of columnar cells with clear cytoplasm. The first prominent theory involves seeding of ovarian tissue into the retroperitoneal space. The inciting event for this seeding is unknown to this day. Support for this theory comes in the form of immunochemical staining matching tissue to cytokeratin 7 and 20 which are also seen in the ovarian type of mucinous cystadenomas. However, this was only verified in a few cases. There are also at least two reported cases of PRMC in a male patient. The other predominant theory suggests that there is a teratoma metaplasia that occurs prior to the formation of the PRMC. The pluripotent cells make for mucinous cystadenomas and other malignant

transformations of this tumor in the retroperitoneum. Review of the literature seems to provide more favor toward the second theory. Both theories however shed light on the path to malignancy PRMC can easily take. This makes diagnosis and removal very important even in the case of benign histology. Therefore, PRMC should be included in the differential for retroperitoneal masses. The rate of recurrence and infection in masses that are not removed is reported as high. The follow-up for these cases is again a controversy due to the lack of case evidence. The treatment of choice is surgical resection [7]. Complete removal is almost always advised when possible. Surgery seems to decrease if not eliminate the incidence of recurrence and infection as well as transformation to malignancy. Two cases of benign PRMC have shown no recurrence or malignant transformation 8 months and 2 years after excision respectively [8,9]. Some other reports have called for sampling or the surrounding organs [10]. In the case reported here a recurrence after drainage was followed by a laparoscopic excision of the tumor. This minimally invasive procedure followed by pathologic identification of the mass was so far successful treatment.

A written HIPAA authorization and informed consent were obtained from the patient on admission to the hospital and confirmed in the office follow-up with an attending physician present.

Case Presentation

A reproductive aged woman Gravida 3 Para 3 came into the general surgeon's office with generalized abdominal pain and feelings of abdominal fullness. Physical exam showed an obese woman with slight tenderness to palpation of the abdomen without guarding or rebound tenderness. Pregnancy and sexual transmitted infection panels were negative. There was no pertinent family history. The patient had a personal history of diabetes. No hospitalizations other than for the un-complicated vaginal births of her children. She had surgical history 5 years ago of a percutaneous drainage of a retroperitoneal cystic mass diagnosed *via*. Magnetic Resonance Imaging (MRI). The patient chose the percutaneous drainage as it was the least invasive method offered. This was in-between child 2 and 3. The fluid was reported as mucinous and non-bloody. The fluid had not been sent to the lab at that time. She had similar symptoms then and now. A computerized tomography image confirmed the

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***Corresponding author:** Manisha E Kotay, American University of the Caribbean School of Medicine (AUC), Sint Maarten, 650 Norfolk St, Dunedin, Florida-34698, USA, Tel: 727-366-4789; E-mail: manishakotay@students.aucmed.edu

retroperitoneal mass had recurred. Since there had been a recurrence before the surgeon decided this time the entire mass should be excised. The surgery was completed laparoscopically. The cyst was drained with a needle as the risk of rupture was determined as high given the masses size and location. Then the walls of the mass were excised and removed in an endobag. The mass was sent to pathology who determined the diagnosis - a primary retroperitoneal mucinous cystadenoma. This was the diagnosis based on the structure of a thin walled mucinous mass consisting of columnar cells. The mass was determined to be benign, so no further treatment was indicated. The woman was advised to follow up in the office for routine post-surgical management and to continue with her life as normal.

Discussion

Here we see a constellation of nonspecific symptoms and definitive mass on imaging. The first treatment given with percutaneous drainage proved not enough to prevent recurrence. It would have been wise to biopsy the mass earlier, as there is a proven path here for malignant transformation. The patient noted above was lucky to have no signs of transformation and needed no further treatment. Over 12 months later, the patient shows no signs of recurrence. It is important to diagnose PRMC because malignancy and infection may occur. In benign cases removal is indicated and likely curative. The literature review shows favour for the theory of pluri-potent cells implanting and creating mucinous cystadenomas in the retroperitoneal space. This theory and the competing theory of seeding from ovarian tissue both show a very easy pathway for malignant transformations of this tumor in the retroperitoneum. It is easy to see why this diagnosis is an important one to consider and an important one to treat with removal as oppose to drainage alone.

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