Primary Retroperitoneal Mucinous Tumor – A Cyst That Is Not So Simple

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Background

- Primary retroperitoneal mucinous tumors are very rare, with few case-reports and case-studies written regarding this neoplasm. Occurring mostly in reproductive aged women, these slow-growing tumors largely go unnoticed until reaching an appreciable size. Clinical examination or self-palpation of a mass-like structure upon abdominal exam is how many primary retroperitoneal mucinous tumors are initially identified. Pathogenesis of these neoplasm remains unknown, as epithelial tissue does not exist in the retroperitoneum. Diagnosis and treatment is obtained through surgical resection. This is a case-report of a patient with pathologic diagnosis of a primary retroperitoneal mucinous tumor.

Case Description

34-year-old female with a past medical history of Crohn's disease, and rheumatoid arthritis on methotrexate has had chronic gastrointestinal symptoms, specifically troubling for nausea and diarrhea. The patient admits her weight is stable and her oral intake is stable.

To evaluate possible causes of her abdominal symptoms, an abdominal CT scan was ordered which showed nephrolithiasis and a simple renal cyst measuring 7 cm that is exophytic cyst off her right kidney (figure 1).

Five years later, the patient developed abdominal pain after physical exercise and noticed an area of swelling and a palpable mass in the right abdomen during a self-preformed abdominal exam. A repeat abdominal CT scan redemonstrated the renal cyst, however larger at this time (figure 2).

An ultrasound was preformed to further evaluate the mass which revealed an 11.4cm right renal mass (figure 3).

It was then recommended that the patient undergo a robotic assisted laparoscopic right renal cyst decortication.

Surgical resection of the mass was successful, and the specimen was sent for pathologic evaluation. Pathologic testing was preformed which initially demonstrated malignant adenocarcinoma cells suggestive of a possible upper gastrointestinal primary tumor.

Pathology slides were examined and identified a primary retroperitoneal mucinous tumor. The tumor margins from the excision were negative.

The patient was then referred to medical oncology for further surveillance and consideration of chemotherapy and for evaluation for further surgical exploration for possible primary tumor identification.

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Images

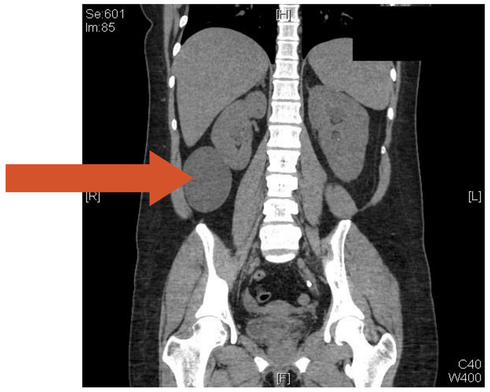

Figure 1: CT scan showing nephrolithiasis and a simple renal cyst measuring 7 cm that is exophytic cyst off her right kidney.

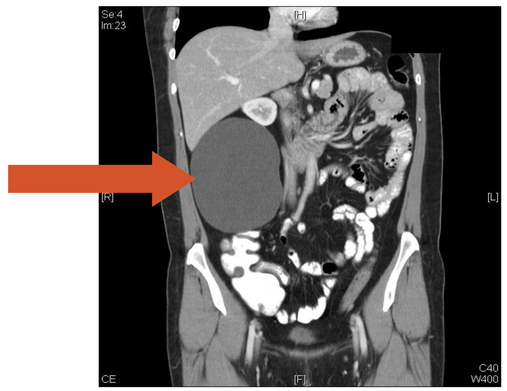

Figure 2: A second abdominal CT scan demonstrated the renal cyst, however larger at this time at 10.4cm in diameter.

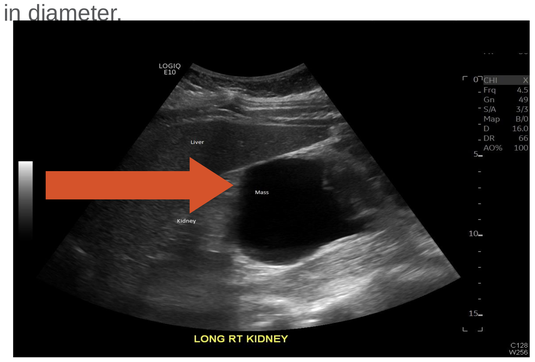

Figure 3: An ultrasound was preformed to further evaluate the mass on 04/29/21, which revealed an 11.4cm right renal cyst.

Discussion

- Primary retroperitoneal mucinous tumors are very rare, occurring in roughly 0.1%-0.2% of all malignancies. Most of these rare tumors are initially identified with abdominal imaging. With 70-80% of all primary retroperitoneal neoplasms being potentially malignant, surgical resection and pathologic evaluation are necessary for diagnosis of the underlying mass. As seen in our patient, the mass was continuing to enlarge prompting surgical removal.
- Once pathologic evaluation was preformed, initial results indicated a possible metastatic lesion. Two distinct morphologies were identified on histology. One, demonstrated mucinous epithelium in a signet ring morphology. While the second, showed more solid and highly anaplastic cells with atypical mitotic figures and a dense fibrotic response.
- Our patient underwent robotic assisted laparoscopic resection of the primary retroperitoneal mucinous tumor with negative margins. The patient tolerated the procedure well and has had no complications post-operatively. Negative tumor markers for pancreatic and colon neoplasm make translocation of anaplastic tissue unlikely in our patient. The exact histogenesis of primary retroperitoneal mucinous tumors continues to remain unclear.
- Standard of care for treatment remains controversial. Primary tumor resection remains agreed upon. However, further surgical resection following tumor removal and chemotherapy remain controversial.

Conclusion

- Primary retroperitoneal mucinous tumors are exceptionally rare. These tumors are largely benign; surgical resection and pathologic evaluation are required for diagnosis. Apart from surgical resection of the tumor, further treatment including further surgical resections and chemotherapy remain controversial. Primary retroperitoneal mucinous tumors should be considered in the differential of a retroperitoneal mass.

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