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Case Report

Upper Abdominal Primary Retroperitoneal Cyst with Unusual Urothelial Histogenesis - An Uncommon Presentation Masquerading as a Hepatic Cyst

Noor Marji, Brett Baskovich, Ahmad Alkhasawneh, Civan Altunkaynak, Ziad Awad and Arun Gopinath*

Department of Pathology, University of Florida college of Medicine Jacksonville Jacksonville, FL, USA

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ABSTRACT

Background: Benign retroperitoneal cysts are uncommon. There is a paucity of literature on primary upper abdominal retroperitoneal cysts of urothelial histogenesis. We report an uncommon presentation of urothelial cyst clinically mimicking a hepatic cyst.

Case presentation: A 21-year old female patient was admitted with intermittent abdominal pain of 9 months duration diagnosed on radiology as a right hepatic cyst. Laparoscopic resection and pathologic examination revealed a primary retroperitoneal cyst with a urothelial histogenesis. This is an unusual clinical presentation of a rare lesion mimicking a hepatic cyst.

Conclusion: This case highlights the unusual anatomic location and the rare occurrence of an upper abdominal retroperitoneal urothelial cyst.

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Background

The retroperitoneum is home to a wide range of pathologic entities. Benign retroperitoneal cysts are uncommon, with a reported average incidence of one in 10500 patients [1]. Primary benign retroperitoneal cysts are rare, and most of the urogenital cysts reported are of Müllerian origin. The urothelium-lined cyst has been rarely reported in the literature and usually occurs in the peri-renal area or near the lower urinary tract. We report an unusual occurrence of a retroperitoneal cyst in a young woman who presented with abdominal pain and underwent surgical resection.

Case Presentation

A 21-year-old Caucasian female with no significant past medical history presented with intermittent abdominal pain for 9 months, for which she was seen at the emergency department several times. An MRI scan (Figure 1) revealed a 3.5 x 6.0 x 5.0 cm (AP X TRV X CC) T2 hyperintense lobulated cystic structure demonstrating some internal

septations and apparently arising from the posterior right hepatic lobe. Peripheral calcifications were also noted. The kidneys showed normal enhancement without any hydronephrosis. Laparoscopic surgical resection of the cyst was performed. Intraoperatively, the cyst was located entirely in the retroperitoneum, hidden under but not connected to the right lobe of the liver, and completely separated from the other abdominal viscera. The cyst fluid was aspirated, and the cyst wall was completely resected, including its focal attachment to the diaphragm.

The macroscopic examination of the specimen revealed a 6.0 x 5.0 x 0.6 cm purple cystic structure in a collapsed state. It had a partly hemorrhagic but smooth inner lining, with patchy areas of calcification. Microscopic examination showed a cyst wall consisting of fibrous tissue with an outer layer of skeletal muscle fibers and adipose tissue. The inner lining consisted of a stratified benign-appearing epithelium (Figures 2a & 2b). The immunohistochemical stains were consistent with urothelial/transitional epithelium (Table 1; Figures 3a & 3b). The patient is now asymptomatic on six months of follow up.

*Correspondence to: Arun Gopinath, Department of Pathology, University of Florida college of Medicine Jacksonville Jacksonville, FL-32209, USA; E-mail: Arun.Gopinath@jax.ufl.edu

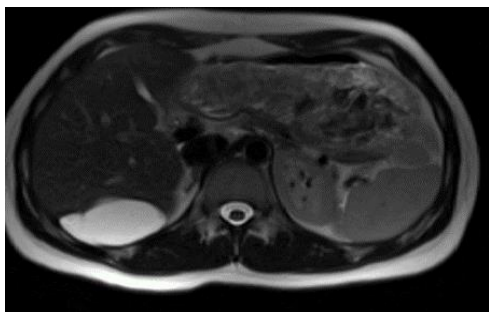


Figure 1: T2-weighted MRI of the abdomen reveals a hyperintense lobulated cystic structure posterior to the right hepatic lobe.

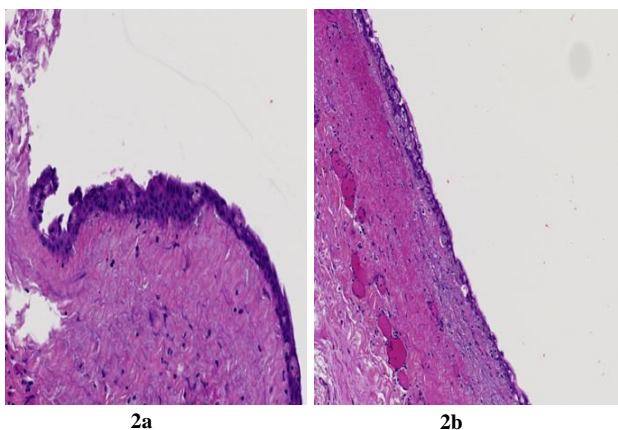


Figure 2a & 2b: The cyst lined by benign-appearing stratified epithelium.

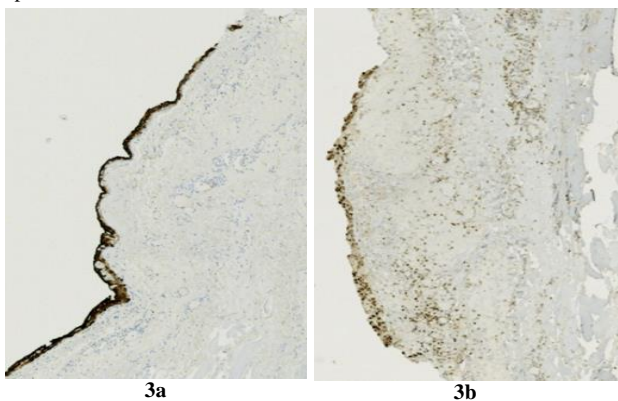


Figure 3a & 3b: Cyst lining diffusely expressing cytokeratin 7 and GATA 3.

Table 1: Results of immunohistochemical studies.

Positive	Negative
CK7	TTF-1
CK20 (umbrella cells)	PAX-8
GATA-3	Calretinin

Discussion

Retroperitoneal cysts are those located in the retroperitoneum with no connection to any anatomic structures [2]. They can be divided into true cysts with an epithelial lining, and pseudocysts without a true epithelial lining. The true cysts are further classified based on embryologic origin and histologic differentiation into lymphatic (lymphangioma),

bronchogenic, enteric, mesothelial, and urogenital cysts [2]. Lymphangiomas are congenital benign cystic masses with endothelial lining that occur due to abnormal development of the lymphatic channels. Most commonly, they occur in the head and neck, and they are unusual in the retroperitoneum [3, 4]. Mesothelial cysts are cystic lesions that originate in the serous lining of body cavities or pelvic viscera. They have no relation to asbestos exposure and occur more frequently in women. Histologically, they can be unilocular or multilocular cysts and contain watery secretions [5]. Urogenital cysts are extremely rare and are considered to originate from vestiges of the embryonic urogenital apparatus. They are classified based on the embryological lines into pronephric, mesonephric, metanephric and Müllerian cysts [6]. They occur near the kidney, behind the colon, and near the head or tail of the pancreas. Most are Müllerian in origin, and they are seen more often in females, usually presenting in adolescence or mid-adult life [6-8].

They have a smooth, glistening lining membrane, are unilocular, and contain a clear serous fluid. Histologically, the cyst wall is thin, consisting of a cellular fibrous tissue lined by a low columnar, cuboidal or flattened epithelium [8]. Müllerian cysts are lined mainly by cuboidal and columnar epithelium with cilia. Though they are considered developmental, they may grow in response to hormonal stimuli [6]. Enteric cysts arise from the posterior foregut, which is the embryologic precursor of the upper gastrointestinal tract. They are lined by the esophageal, gastric, or intestinal mucosa and typically occur in the posterior mediastinum. Tailgut cysts arise from the vestiges of the embryonic hindgut and occur between the rectum and sacrum. Most reported lesions have been benign, but they can undergo malignant transformation into adenocarcinoma [5]. The tailgut cysts are usually multicystic and lined by different types of epithelium, including ciliated or mucin-secreting columnar, transitional, and squamous [9].

In our patient, the cyst was multiloculated with epithelium most consistent with transitional epithelium (urothelium). This was strongly supported by positive immunohistochemical expression of CK7 (Figure 3a), CK20 (umbrella cells), and GATA-3 (Figure 3b), with negative PAX8 (Müllerian marker), TTF-1 and calretinin (mesothelial marker). Given the epithelial (transitional) nature of the cyst lining, the retroperitoneal location, and the immunohistochemical pattern, the other retroperitoneal cysts (lymphangioma), enteric, mesothelial, and urogenital) were excluded. Tailgut cysts can rarely be lined by transitional epithelium/urothelium. However, in this patient, the cyst was in the upper abdomen behind the right lobe of the liver, compared to the usual retrosacral location of the tailgut cyst. Although this lesion may fall under the umbrella of urogenital cysts, the site of occurrence is uncommon. Even though rare case reports of primary transitional carcinoma of the retroperitoneum have been published, an extensive search has not revealed any documentation of primary upper abdominal retroperitoneal urothelial cyst [10-12]. Some possible causes of its development include origin from a cystic diverticulum with complete detachment from the urinary tract, embryonic urogenital remnants, coelomic metaplasia, and monodermal teratoma.

Conclusion

We present an unusual occurrence of a retroperitoneal urothelial cyst in a patient presenting with abdominal pain, which masqueraded as a

hepatic cyst on imaging. The site of occurrence outside the renal/perirenal area makes this a unique case. Further discussion and studies with adequate follow up are needed regarding its categorization and nomenclature. Additionally, rare reports of primary retroperitoneal urothelial/transitional carcinomas in the literature warrant additional follow-up for these patients and consideration of the possibility of aggressive transformation.

Conflicts of Interest

None.

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