Giant Mesenteric Cyst: A Case Report And Review Of The Literature

L Pantanowitz, M Botero

Abstract
We present a case of a giant mesenteric cyst successfully removed surgically. The rarity of such mesenteric cysts makes them difficult to diagnose clinically and pathologically. Simple mesenteric cysts need to be distinguished from lymphangiomas, hemangiomas, pancreatic pseudocysts, endometriomas, loculated ascites, peritoneal inclusion cysts (cystic mesothelioma), cystic mesenteric panniculitis, hydatid cysts, cystic teratomas and urogenital cysts of the mesentery. Complete enucleation of these cysts is considered the procedure of choice, to prevent complications, recurrence and possible malignant transformation.

CASE REPORT
A 39-year-old otherwise healthy man presented to hospital because of an increasing abdominal girth, which he attributed to merely getting fat. He experienced abdominal discomfort only late in his presentation. On examination, his abdomen was significantly distended, non-tender to palpation and dull to percussion. A fluid thrill could be elicited through his abdomen. A computerized tomographic (CT) scan revealed a large intra-abdominal cyst, with thin intracystic septations (figure 1). The cyst had displaced his bowel throughout the entire abdomen and extended inferiorly into his pelvis. Intraoperatively, it was found to be intimately associated with his mesentery. Grossly, the multicystic mass was tan in color, measured 23 x 15 x 3 cm and contained approximately five liters of serous fluid. There were no papillary excrescences or areas of hemorrhage and necrosis identified. Microscopically, the fibrous wall contained sparse, scattered reactive lymphocytes and was lined focally by cuboidal to attenuated epithelium. The epithelial cells were immunoreactive with low- and high-molecular weight cytokeratins, but failed to stain with factor VIII (figure 2).

Figure 1
Figure 1: Abdominal CT scan demonstrating the large multicystic mass with fine septations.
DISCUSSION

Mesenteric cysts are one of the most rare intra-abdominal tumors. The reported incidence ranges from 1/20,000 to 1/250,000 admissions to hospital. Their rarity makes them difficult to diagnose both clinically and pathologically. They are also poorly understood clinical entities that are difficult to classify. Benevieni, an Italian anatomist, was the first person to detect a mesenteric cyst in 1507, while performing an autopsy on an 8-year-old boy. Cysts that are present in the mesentery, omentum and retroperitoneum are comparable embryologically and pathologically. These anatomical structures are all lined by peritoneum and composed of connective tissue containing fat, lymphatic channels, blood vessels, scant muscle fibers and neural tissue. Therefore, cysts arising in any of these locations may originate from any of these different tissues. It has been suggested that mesenteric cysts develop when the mesenteric leaves fail to fuse during development. They are also thought to arise following localized degeneration of lymph nodes or from lymphatics that are congenitally malformed, malpositioned or obstructed. Mesenteric cysts have now been categorized into four etiologic groups: (i) embryonic and developmental; (ii) traumatic and acquired; (iii) infective and degenerative; and (iv) neoplastic cysts.

Mesenteric cysts are most common in the fourth decade of life, but may also affect young children. They appear to have no significant gender or race predilection. Cysts are most commonly located within the mesentery of the ileum, followed by the omentum, mesocolon and retroperitoneum. The occurrence of multiple cysts is not uncommon. Cysts can be unilocular or multilocular, and may contain chylous, serous or infrequently hemorrhagic fluid. Mesenteric cysts filled with milk of calcium have been described, and there have also been rare reports of gas accumulation within cysts. The cysts can remain asymptomatic and therefore grow to giant proportions, as illustrated in the present case. They may be mistaken for ascites on physical examination and abdominal imaging studies. Patients may complain of chronic abdominal complaints, or exceptionally present with an acute abdomen. Complications include torsion, infarction, volvulus formation, perforation, infection, anemia from intracystic hemorrhage, intestinal obstruction and obstructive uropathy. Children are most likely to develop life-threatening complications. Mesenteric cysts can also develop into either carcinoma or sarcoma. Cystic abdominal masses are easily evaluated radiologically, obviating the need for more invasive studies. Information regarding their location, size, origin and internal structure may aid in the differential diagnosis.

The differential diagnosis for cystic lesions in the mesentery includes lymphangioma, pancreatic pseudocysts, hemangioma, endometriosis, loculated ascites (usually tuberculous), peritoneal inclusion cysts, cystic mesenteric panniculitis (sclerosing mesenteritis), hydatid cyst, cystic teratoma and urogenital cysts. Simple mesenteric cysts have a fibrous wall that is devoid of a defined muscular layer. They are lined by an attenuated layer of epithelium. Those without an epithelial lining are thought to be of traumatic origin. Calcification and reactive, chronic inflammatory changes may be present within their fibrous walls. The lining epithelium of cystic lymphangiomas is also flat or fusiform, but is immunoreactive for endothelial markers. Duplication anomalies (enterogenous cysts) that are lined by intestinal epithelium may communicate with, as well as share a blood supply with, the gastrointestinal tract. They contain a more well defined muscular layer that contains myenteric neural elements. Urogenital cysts of the mesentery are lined by more cuboidal to columnar epithelium, often with distinct cilia. Urogenital cysts with primitive renal structures incorporated into their cyst wall are probably of mesonephric derivation. Mesenteric cysts derived from implanted ovarian tissue, known as the mesenteric cyst-ovarian implant (or remnant) syndrome, usually arise in patients who have had previous oophorectomies. These cysts are lined by luteal cells, and normal ovarian tissue may be located adjacent to the cyst wall. The wall of an echinococcal (hydatid) cyst comprises an external, anucleate and amphophilic laminated layer that is about 1 mm thick, surrounded by another thin outer layer of fibrous tissue produced by the host. A germinal membrane that is only a
single cell layer thick may line this laminated layer. Sterile cysts lack a germinal membrane. Only fertile hydatid cysts therefore contain pathognomonic brood capsules, which develop on stalks from this germinal layer. Detached free-floating daughter cysts, which rupture to release scolices, are referred to as “hydatid sand.”

Peritoneal inclusion cysts (so-called cystic mesothelioma) usually occur in women of reproductive age. Mesothelial cysts may be arranged in grape-like clusters or present as isolated lesions studding the surface of the peritoneum. They often contain more mucinous or gelatinous fluid. Microscopically, they are lined by a single layer of cuboidal or flattened mesothelium that may focally form papillary projections, nests or tubules. Occasionally this undergoes squamous metaplasia. There may be mild to moderate cytologic atypia, but no mitoses. The formation of glands that have an infiltrative pattern may be mistaken for cancer. Vacuolated mesothelial cells may easily resemble signet-ring carcinoma. Unlike other cysts of the mesentery and retroperitoneum, cystic mesothelioma has a propensity to recur in up to 50% of cases. Rare transformation to malignant mesothelioma, usually of low grade, has been reported. In these cases the cysts are lined, at least focally, by more atypical mesothelial cells. In mesenteric panniculitis (or sclerosing mesenteritis) a marked inflammatory process results in the formation of multiple pseudocysts, which may mimic cystic mesothelioma. The pseudocysts in this condition, however, contain a more marked chronic inflammatory process composed of reactive lymphocytes and plasma cells, associated with lymphedema and fat necrosis. If mesothelioma is a diagnostic concern, a definitive answer can be achieved using histochemical and immunohistochemical stains, as well as electron microscopy.

Surgery is the mainstay of treatment, and the only definitive diagnostic modality for simple mesenteric cysts. Aspiration of the cyst alone should not be performed. Complete enucleation of cysts is considered to be the procedure of choice, to prevent recurrence and possible malignant transformation. Segmental intestinal resection may be necessary if the adjacent bowel has a compromised blood supply. For multiple cysts or those technically difficult to excise completely, such as those located within the retroperitoneum, marsupialization with careful follow-up may be necessary. Finally, there are reports of successful laparoscopic resection of mesenteric cysts.

FOR CORRESPONDENCE
Liron Pantanowitz, MD
Chief Resident in Pathology
Tel: 617-667-4344
Fax: 617-667-7120
E-mail: lpantano@caregroup.harvard.edu

References
Author Information

Liron Pantanowitz, MD
Chief Resident in Pathology, Department of Pathology, Anatomical & Clinical Pathology, Beth Israel Deaconess Medical Center

Maria Botero, MD
Resident in Pathology, Department of Pathology, Anatomical Pathology, Beth Israel Deaconess Medical Center