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Review article

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MESENTERIC CYSTS

SUMMARY

Mesenteric cysts are rare intraabdominal tumors usually briefly reported in textbooks. Their low incidence is the main reason for the lack of simple and clinically relevant classification of these masses. The aim of the study was to identify and characterize different groups of mesenteric cysts and to state their most distinguished features concerning clinical presentation, histopathological structure, diagnosis and treatment by a literature review and present the most appropriate classification available in literature. Classification based essentially on histopathological features seems to be most comprehensive and includes the six following groups of mesenteric cysts: 1) cysts of lymphatic origin, 2) cysts of mesothelial origin, 3) cysts of enteric origin, 4) cysts of urogenital origin, 5) mature cystic teratoma and 6) pseudocysts. Surgical extirpation of mesenteric cyst with resection of adjacent organs if necessary is mandatory in order to exclude malignant alteration and prevent complications such as inflammation, hemorrhage, torsion or rupture. The preferred mode of treatment is enucleation.

Keywords: **mesenteric cyst, classification, treatment**

INTRODUCTION

Mesenteric cysts are rare intra-abdominal tumors generally omited or briefly reported in textbooks. They arise with an incidence of 1/105000 hospitalized adult surgical patients and 1/20000 admissions in children (1). Their rarity had fostered a lack of information and difficulty in classification. These cysts may occur in every part of the mesentery, from duodenum to rectum. Most frequently cysts are localized in small bowel mesentery (ileum in 60%) and right mesocolon (ascending colon in 40%). Although some of mesenteric cyst are well-defined (e.g. chylous cysts) (2), there is still a controversy about the etiology and classification of most of these cystic tumors. Many authors consider mesenteric, omental and retroperitoneal cysts as one group of entities, because they derive from the same same embryological structures (3,4), whereas others define mesenteric cysts as cysts arising in or near the mesentery without connection to retroperitoneal organs (1). Mesenteric cysts have simillar pathogenesis, but may have different histopathological origin and structure. Most often they represent ectopic lymphatic tissue – lymphatic, chylous cysts. Lately, electron-microscopic findings brought new elements in the identification of endothelial and mesothelial cells which allowed a new classification of mesenteric cysts in two large groups: those of lymphatic origin (such as simple lymphatic cyst and lymhangioma) and those of mesothelial origin (such as simple mesothelial cyst, benign cystic mesothelioma and malignant cystic mesothelioma). Since the incidence and mode of presentation significantly

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differ between these two most frequent types of mesenteric cysts, a new classification separating them into distinctive categories is warranted and necessary.

Historical background

Mesenteric cyst was first described in 1507 by Benevieni, Florentine anatomist, during the autopsy on an eight-year-old boy (5). However, it was not until 1842 when Rokitansky gave the first description of a chylous mesenteric cyst (3). In 1880, Tillaux performed the first successful operation (resection) on a cystic mesenteric tumor (2). After him, Pean reported the first marsupialization of a mesenteric cyst in 1883. Since then, the number of cases reported in the literature increased from 100 in 1897 up to about 900 until today, providing more informations on etiology, nature, symptomatology, diagnosis, treatment and prognosis. Still, even today, the literature reports on cystic tumors of mesentery are relatively rare.

Epidemiology

Mesenteric cysts are rare abdominal findings. Mesenteric cysts occur with very small incidence of 1/105000 admissions in adults (1), mainly later in life (fifth decade) and with female predominance in occurrence (twice more frequently)(6). The exception are cystic lymphangiomas which mostly occur in the first decade of life (up to 12 years of age), with incidence of 1/20000 hospitalized children (1,6) and with male predominance (7). Also, although some mesothelial cysts have been reported in children, most of them occur in young and middle-aged women (16,17). This lack of clinical experience in treatment of this rare surgical entities is probably the cause of controversies about their etiopathogenesis and histopathological classification.

Classification

Mesenteric, omental and retroperitoneal cysts derive from the same embryological structures. However, in spite of the similar embryological origin and etiopathogenesis, their histopathological structure is significantly different, which for a long time was the reason for the lack of systematized, clinically useful classification of these formations. There are several suggested classifications of mesenteric cysts based on etiology and cyst content (Caropreso 1974, Moynihan 1987), among which the most accepted and clinically most commonly used is the one proposed by Beahrs in 1950 (8,9,10). This classification is based mainly on etiological and clinical features of mesenteric cysts which are divided into 4 groups: 1) embryonal and developmental, 2) traumatic, 3) neoplastic and 4) infectious. However, this classification significantly disregard histological structure of cysts. As a result, cysts of mesothelial origin were excluded. In 1979, Mennemeyer and Smith provided the foundation for adequate classification of mesenteric cysts (11). They established and clearly defined histological, ultrastructural and especially immunohistochemical features and differences between the cysts of mesothelial and endothelial origin (that is lymphatic cysts) and also between the cysts inside the same histological group. For example, the distinction between simple lymphatic cyst and lymphangioma or simple mesothelial cyst and benign or malignant (multi)cystic mesothelioma is of a great clinical and therapeutic importance because these tumors have different clinical behavior and prognosis. Simple lymphatic and mesothelial cysts usually remain stable and as a rule are asymptomatic over the time, whereas lymphangiomas and benign cystic mesotheliomas may have invasive properties and aggresive evolution. The only genuine malignant tumor in this classification is malignant cystic mesothelioma which may, although rarely, simulate the gross appearance of benign cystic mesothelioma and, therefore, lead to misdiagnosis. Hence, these findings enabled the creation of new widely clinically accepted classification based essentially on histopathological features which included six groups of mesenetric cysts (Table 1).

Table 1. Classification of mesenteric cysts

1.	**Cysts of lymphatic origin**
	1a Simple lymphatic cysts
	1b Lymphangiomas
2.	**Cysts of mesothelial origin**
	2a Simple mesothelial cysts
	2b Benign cystic mesotheliomas
	2c Malignant cystic mesotheliomas
3.	**Cysts of enteric origin**
	3a Enteric duplication cysts
	3b Enteric cysts
4.	**Cysts of urogenital origin**
5.	**Mature cystic teratomas (dermoid cysts)**
6.	**Nonpancreatic pseudocysts**
	6a Cysts of traumatic origin
	6b Cysts of infectious origin

Cysts of lymphatic origin have the wall containing smooth muscle fibers, lymphoid tissue and/or occasional foam cells and are characteristically lined by flat endothelial cells. In contrast,

cysts of mesothelial origin are lined by flat, cuboidal or columnar cells and their wall is fibrous without any lymphatic structures. Precise distinction between endothelial and mesothelial cells and their further characterization can be achieved by immunohistological analysis (Table 2).

and usually have a thick fibrous wall without an inner epithelial lining (Figure 1). Therefore, such cysts may be considered as nonpancreatic pseudocysts.

Table 2. Differential diagnosis criteria between cysts of mesothelial and lymphatic origin

	Mesothelial cysts	**Lymphatic cysts**
Citology	rounded cells	elongated cells
	regular, round nuclei	oval, oblong nuclei
	prominent, single nucleoli	small nucleoli
	abundant cytoplasm	scant cytoplasm
Histology	uni-/multiloculated	uni-/multiloculated
	flattened, cuboidal or columnar lining cells	flattened lining cells
	fibrous wall	the wall contains smooth muscle, lymphoid tissue and rarely foam cells
Immunohistology		
Factor VIII	-	+
CD31	-	+
Keratin total	+	-
Vimentine	+	+
EMA	+	+

The distinction between malignant and benign cystic mesothelioma is based on the degree of cellular proliferation, atypia and microcystic changes and usually is not difficult. Cysts of enteric origin are easily recognized by their enteric mucosal lining. Enteric duplication cysts are true diverticulae of the bowel pinched off during gestation and contain an enteric mucosa, a muscle layer and a nervous plexus (8). Enteric cysts, in contrast, result from the migration of an acquired diverticula into the mesentery and are lined by an enteric mucosa only, without muscle layer and nervous plexus. Mature cystic teratomas usually contain mucoid fluid and are lined by an epithelium. Also, these tumors characteristically contain extraabdominal tissue, such as skin and skin appendages, cartilage, bone, teeth or adipose tissue (12). Urogenital cysts are rare mesenteric cysts derived from vestigial remnants of the urogenital apparatus such as the Wolffian and Müllerian ducts and their lining cells have a transitional, squamoid or clear-cell appearance and their wall contains well-developed smoth muscle fibers. Their further distinctive embryological subclassification into pronephric, mesonephric, metanephric or Müllerian type is possible, but often difficult (13). Traumatic and infectious pseudocysts are histologically similar to pancreatic pseudocyst

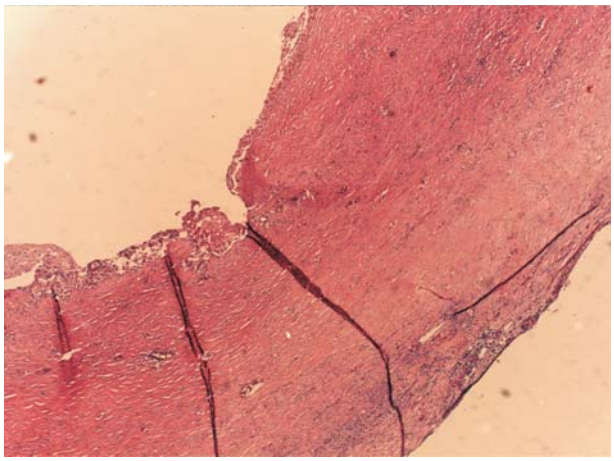

Figure 1. Traumatic cyst of mesentery complicated with chronic inflammation (thick fibrous wall of the cyst imbued with chronic inflammatory cells, with the presence of multiple aggregates of foam macrophages with hemosiderin, the presence of cholesterin granuloma on the cyst's wall and the absence of epithel)

Etiology

The etiology of mesenteric cysts is various. Simple lymphatic and mesothelial cysts are most likely congenital, while the origin of lymphangiomas and benign cystic mesotheliomas is not yet clear

(14). Some authors favor their developmental origin (15), whereas others consider them as neoplastic formations (2). It was established that the occurrence of benign cystic mesotheliomas is frequently associated with a history of previous pelvic inflammatory processes and/or surgery and endometriosis (16).

Clinical presentation

Mesenteric cysts are mostly asymptomatic and, if present symptoms, are quite unspecific. Nevertheless, according to Vanek, mesenteric cysts may be presented with chronic vague abdominal pain, painless abdominal mass or even with acute abdomen (1). Later, it is more frequent in cases of complicated mesenteric cysts (inflammation, abscess, rupture). Contrary to adults, in children, mesenteric cysts become symptomatically very frequent, especially lymphangiomas (17). The most common physical finding (in 61% of patients) is compressible soft abdominal mass, relatively movable transversely (but not longitudinally) which corresponds to the mobility of mesentery (1,8). The occurrence and character of the symptoms depend on the position and size and only partially on the type of mesenteric cyst. For example, the localization and mode of presentation seem to differ between simple mesothelial cysts and benign cystic mesotheliomas. The former are usually reported within the mesentery and remain asymptomatic (17), whereas the latter have been frequently observed within the pelvis and usually become symptomatic. Symptoms are most frequently caused by the compressive effect of the cyst on surrounding structures and rarely by complications of the cyst: inflammation, abscess, rupture. Contrary to simple lymphatic and mesothelial cysts, lymphangiomas and benign cystic mesotheliomas become symptomatic more often over time because of progressive enlargement (17). The size of cyst and age of patient influence the clinical presentation. In children, mesenteric cysts typically clinically present with acute abdomen which may simulate acute appendicitis, intestinal torsion or invagination, while adults have more variable, unspecific and indolent symptoms that include vague abdominal pain (82%), nausea and vomiting (45%), constipation (27%) and diarrhea (6%)(1,2,8). In cases of inflammatory and/or purulent complications and rupture mesenteric cysts may have a clinical presentation of circumscript or diffuse peritonitis, that is acute abdomen and septic shock.

Diagnosis

Laboratory investigation usually does not yield any significant information. Nevertheless, a precise preoperative diagnosis usually can be established with thorough physical examination and radiographic investigation. US and CT of the abdomen can distinguish between solid and cystic character of abdominal mass and determine its size, position, extension, relation to surrounding structures and features of cyst's wall and content (thickness of the wall, presence of septations, calcifications and/or fat, density of liquid content, presence of fluid-fluid levels or echogenic debris resulting from an old internal bleeding or hematoma etc.)(Figure 2). It is rarely necessary to perform additional diagnostic procedures that may help differentiate between cystic and solid tumor and further characterize the cyst, like NMR, fine needle aspiration and cytological analysis and explorative laparoscopy.

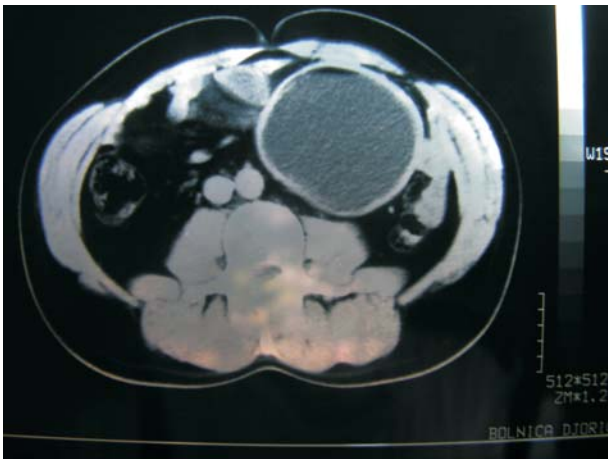

Figure 2. CT scan presenting mesenteric cyst

Treatment

In cases of large mesenteric cysts, especially symptomatic, surgical extirpation is mandatory in order to exclude malignant alteration (such as in retrorectal teratomas) and prevent complications such as inflammation, hemorrhage, torsion or rupture. The distinction between simple lymphatic cysts and lymphangiomas or between simple mesothelial cysts and benign cystic mesotheliomas can be achieved intraoperatively based on macroscopic characteristics. Simple cysts are usually small (1-5cm in diameter) and unilocular, whereas lymphangiomas and benign cystic mesotheliomas are large and include multiple cysts of several size. Still, occasionally their gross appearance may be similar and the distinction is not

possible. Malignant mesothelioma may, in rare cases, have grossly visible cystic foci and extensive microcystic changes. Mature cystic teratomas (formerly named dermoid cysts) present multiple solid and cystic areas and extraabdominal tissue is frequently observed (12,18). Urogenital cysts are usually confined to the adnexal region (13) and traumatic and infectious pseudocysts have extraordinary thick fibrous wall. The preferred mode of treatment is enucleation of mesenteric cyst (1), that is atraumatic separation of the cyst from surrounding leaves of mesentery (Figure 3). However, sometimes enucleation can not be performed safely because of firm adhesions of the cyst wall to surrounding mesenteric tissue and/or other structures. This is mostly the case with lymphangiomas and benign cystic mesotheliomas which can strongly adhere to surrounding vital structures and impede or disable their safe extirpation. Contrary, enucleation of simple lymphatic and mesothelial cysts is usually easily feasible. In order to perform complete excision of these cysts, a resection of adjacent organs may occasionally be necessary (bowel, spleen, pancreatic tail). A bowel resection is necessary in only 1/3 of adults, but becomes necessary in up to 50-60% children with mesenteric cyst (7).

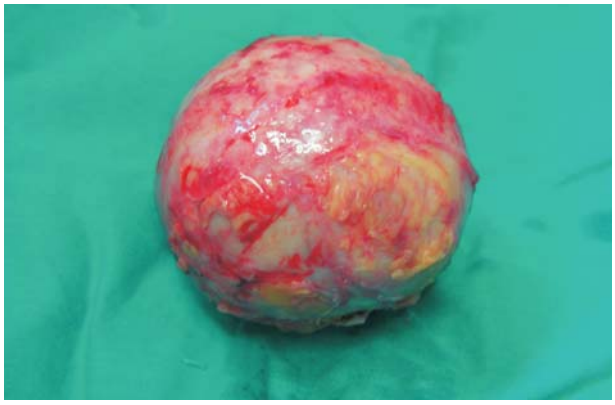

Figure 3. Enucleated mesenteric cyst

Prognosis

With the exception of malignant cystic mesothelioma, all mesenteric cysts are benign and their total excision is usually curative with very good prognosis. However, benign cystic mesotheliomas and lymphangiomas have a high tendency to recur if not resected completely. No metastatic potential has ever been demonstrated with benign cystic mesotheliomas or lymphangiomas. Nevertheless, several deaths from progressive local enlargement have been reported (7,18). Malignant transformation of mesenteric cyst has been suspected in four cases and truly objectivated only in one of them (malignant transformation of previously benign enteric cyst)(19).

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MEZENTERIČNE CISTE

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SAŽETAK

Ciste mezenterijuma su retki intraabdominalni tumori, obično ukratko predstavljeni u udžbenicima. Njihova mala incidenca je osnovni razlog nedostatka jednostavne i klinički relevantne klasifikacije ovih tumora. Cilj rada bio je da pregledom literature identifikujemo različite grupe mezenteričnih cista i istaknemo njihove najznačajnije karakteristike u pogledu kliničke prezentacije, histopatološke strukture, dijagnoze i lečenje i da predstavimo najadekvatniju klasifikaciju dostupnu u literaturi. Podela koja se bazira na histopatološkim karakteristikama čini se najsveobuhvatnijom i uključuje sledećih 6 grupa mezenteričnih cista: 1. ciste limfatičnog porekla, 2. ciste mezotelnog porekla, 3. ciste enteričnog porekla, 4. ciste urogenitalnog porekla, 5. zreli cistični teratom i 6. pseudociste. Hirurška ekstirpacija mezenteričnih cista sa resekcijom adheriranih organa, ukoliko je neophodno, obavezna je u cilju isključenja maligne alteracije i prevencije komplikacija kao što su zapaljenje, krvarenje, torzija ili ruptura. Poželjan način operativnog lečenja je enukleacija.

Ključne reči: **mezenterična cista, klasifikacija, lečenje**