Extraovarian mature cystic teratoma of the mesentery. A case report and literature review

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Summary

A case is reported of a 60-year old woman who had intermittent abdominal pain due to a mesenteric teratoma which was misdiagnosed by the standard methods of abdominal tumor diagnosis as an ovarian tumor. The neoplasm measured 9 x 8 x 8 cm, was filled by hair and sebum, and histologically presented the typical features of a mature cystic teratoma. The mesentery and overlying small intestine showed an extensive inflammatory granulomatous reaction. The uterus and adnexa were free of neoplasmatic disease. This case of extra ovarian mature cystic teratoma which developed in the mesentery is unique among > 2000 ovarian tumors examined during a 30-year period at Aretaieion Hospital Pathology Laboratory.

Key words: Mesentery; Teratoma; Dermoid cyst.

Introduction

Teratomas (dermoid cysts) of the mesentery are the rarest of all mesenteric cystic tumors [1]. Mesenteric cysts and cystic mesenteric tumors are very rare abdominal growths. They are usually located in the ileum and right colon mesentery but rare cases have been reported in other places from the duodenum to the rectum [2].

There are several suggested classifications of cystic mesenteric tumors but the classification based on histopathologic features is most commonly used [3] and classes the cysts as:

a) cysts of lymphatic origin and cystic lymphangiomas,

b) cysts of mesothelial origin, benign or malignant cystic mesotheliomas

c) enteric cysts,

d) cysts of urogenital origin,

e) dermoid cysts and

f) pseudo cysts, of infectious or traumatic etiology.

Dermoid cysts (mature cystic teratoma) are dysembryogenetic lesions that have an epithelial wall lining and may contain sebum, fatty material, and/or hair [4]. Sometimes a solid wall nodule "Rokitansky dermoid plug", consisting of squamous cells, keratin, and skin adnexa is present, often with calcification [4].

Most cases reported in the literature were mature teratomas which developed in young women [5].

There is evidence that extraovarian dermoids develop in ectodermal inclusions of multipotential cells during closure of embryonal plates or fissures, or at the point of union of ectodermal and other structures [1, 5-7]. While this theory is quite adequate to explain those teratomas arising near the sites of union of embryonic fissures, such as the orbital region, branchial clefts and in the midline of the trunk, it does not explain teratomas arising in the mesentery. There is evidence that clusters of multipotential cells might have been derived from the Wolffian body or the Mullerian tube [1, 8-10].

The aim of this report is to describe the clinicopathological features of this rare abdominal tumor, unique in the archives of the Pathology Laboratory of Aretaieion Hospital.

Case Report

A 60-year-old woman, who suffered from progressive abdominal fullness and intermittent tenderness of the abdomen for a month, was referred to the 2nd Clinic of Obstetrics and Gynecology of Aretaieion University Hospital. The patient's past history and family history were unremarkable. No trauma or previous surgery was reported and the patient did not have nausea, vomiting or changes in bowel habits. Physical examination revealed a soft, yet distended abdomen with a mobile nontender abdominal mass. There were no other abnormal physical findings and routine laboratory studies were normal. Abdominal X-ray showed a soft tissue mass displacing the bowel gas with calcification components.

Ultrasound examination revealed a large cystic mass with well defined borders located in the midline in the mesenterium. The mass was clearly separable from the liver, pancreas, spleen and kidneys. Exploratory laparotomy was performed and a large cystic tumor was found located in the small bowel, proximal to the end of the ileum. Surgical resection of the mesenteric tumor was performed. Partial resection of the ileum (approximately 25 cm in length) was also performed because of the tumor location and the presence of extensive inflammatory changes in the bowel wall.

Gross pathological evaluation showed a tumor consistent with teratoma (Figure 1).

Total hysterectomy with bilateral salpingo-oophorectomy was also performed to exclude the case of a malignant ovarian tumor causing metastatic lesions to the abdomen.

Pathological examination showed a cystic tumor measuring 9 x 8 x 8 cm with a wall thickness of 0.5-0.8 cm filled with hair and sebum. The thicker portion of the wall showed development of skin, hair follicles, skin adnexa, fat, cartilage and bone (Figure 3). The diagnosis of a mature cystic teratoma was confirmed. The specimen of the small bowel and mesentery excised

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Fig. 1

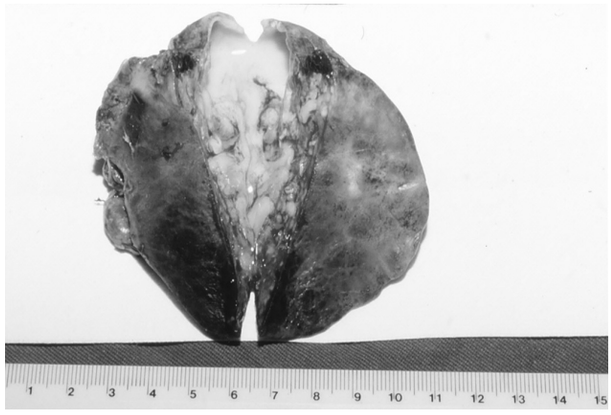

Fig. 2

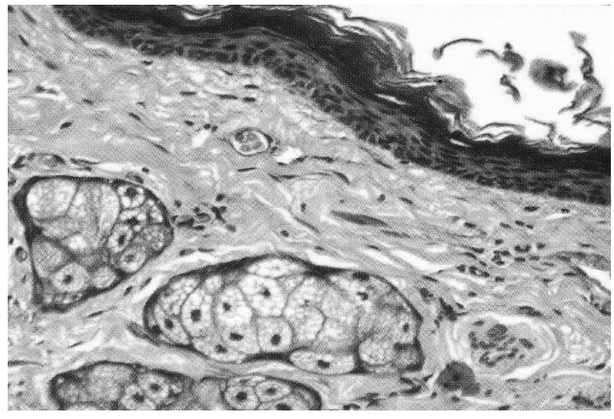

Fig. 3

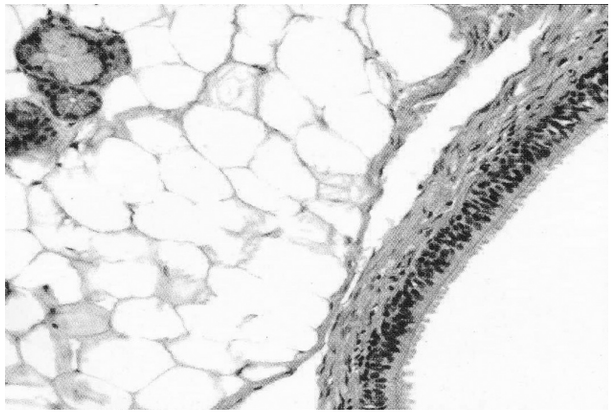

Figure 1. — Gross section of cystic teratoma filled with sebus.

Figure 2. — Histological section showing the epithelial lining of the cystic wall with dermal adnexa (hematoxylin-eosin x 100).

Figure 3. — Histological section showing farty tissue, respiratory epithelium and mucinous glands in the wall of the cystic teratoma (hematoxylin-eosin x 100).

showed extensive inflammatory reaction with granulomas of foreign body nature, probably from a reaction to minute quantities of sebum that escaped from the teratoma.

The patient is well five years after surgery.

Discussion

Teratomas are neoplasms that originate from pluripotent cells and are composed of a wide diversity of tissues, foreign to the organ or anatomic site in which they arise [5]. Benign teratomas are solitary, rounded masses, usually well encapsulated and consisting of both solid and cystic areas [6]. Most mesenteric teratomas are composed of mature tissue elements derived from all three layers. A dermoid cyst is a thin-walled unilocular cystic mass containing sebum. It is lined by a stratified squamous epithelium and contains various embryonic tissues.

Teratomas of the mesentery are very rare and fewer than 25 cases have been reported, mostly as single case reports and occurring mostly in infants, children and young adults [5, 7-11].

Mesenteric teratomas are more common in females with one case reported of a ruptured mesenteric teratoma in a young man [12]. The may arise in any portion of the small bowel mesentery. Most reported cases are of the dermoid cyst type [7-11], presenting a simple epidermal lining of the cyst wall.

The clinical picture of mesenteric tumors is not pathognomonic. A mesenteric cyst should be suspected in the presence of a painless abdominal tumor, with occasionally painful abdominal pressure, normal laboratory findings and good general condition of the patient [2]. There are no cardinal symptoms and all complaints result from purely mechanical disturbances.

The symptoms reported are usually caused by the torsion of the lesion leading to abdominal pain. However a rare case has been reported of a spontaneous intramesenteric rupture [12]. In symptomatic cases, acute or chronic abdominal pain is the most common feature, whereas other symptomatology depends on the location of the tumor, the size and the consequential abdominal organ compression (intestinal obstruction, hydronephrosis, lower extremity lymphedema).

Other symptoms are related to volvulus, intussusception, adhesions and perhaps most rare, hemorrhage from one of the thin walled vessels into the mesentery or the peritoneal cavity [13].

One case associated with autoimmune hemolytic anemia has been reported [8]. The diagnosis should be made by use of all standard methods of abdominal tumor diagnosis, with ultrasonography (US) and computed tomography (CT) [14-21]. Plain radiographs of the abdomen usually demonstrate a large soft tissue mass. Mature cystic teratomas are easily diagnosed based on

imaging studies which reveal their characteristic intratumoral fat and calcification components. US is useful for analyzing the contents of these lesions and must be utilized to search for possible pelvic anomalies, allowing diagnosis of pendunculated ovarian cysts for example. Although mature cystic teratomas show various appearances on US, most can be characterized by the presence of echogenic sebaceous material and calcification. Adipose tissue is seen in 67-75% of the cases and teeth are present in 31% [14].

CT offers better sensitivity for evaluating the effect of such cysts on neighboring viscera and particularly on the intestine. CT is also better for identification of the fatty components indicative of dermoid or chylous cysts [15]. Magnetic resonance imaging is more sensitive for detecting microscopic fat than CT or US [18, 19].

Finally, selective arteriography of the upper mesenteric artery may provide useful information concerning the topography of the tumor in relation to the mesenteral vascular trunk [16].

An exact diagnosis can only be made after open laparotomy, since preoperative diagnosis by even CT is just 25% accurate. Surgery is required to make a definitive diagnosis of mesenteric tumors and to avoid complications [17].

The differential diagnosis of a mesenteric predominantly cystic lesion includes: non neoplastic cysts, such as a pseudocyst, enteric duplication cyst, enteric cyst, mesothelial cyst, dermoid cyst, lymphocele or parasitic cyst and cystic neoplasms, such as cystic lymphangioma, cystic mesothelioma, cystic pancreatic neoplasms and cystic degeneration of solid tumors, such as leiomyoma or leiomyosarcoma. Among these conditions, some may contain a fat component in their cystic portion, including a dermoid cyst, cystic lymphangioma, lymphocele and liposarcoma with cystic degeneration [20]. Fat can be defined on a CT scan. However the definite diagnosis is made by histological examination.

Total cystectomy is the therapeutic method of choice. The open method has been preferred, although reports on successful cystectomy by the laparoscopic method have already appeared in the literature [2]. Because of the slow growth of dermoid cysts (1.8 mm per year) nonsurgical management for asymptomatic lesions less than 6 cm in diameter is recommended [18].

The prognosis of mesenteric teratoma is considered excellent and there is no history of recurrence in any of the reported cases of proved dermoid cysts of the mesentery.

References:

[1] Penberthy G.C., Brownson K.M.: "Dermoid cysts of the mesentery". *Ann. Surg.,* 1937, *107*, 566.

[2] Huis M., Balija M., Lez C., Szerda F., Stulhofer M.: "Mesenteric cysts". *Acta Med. Croatica,* 2002, *56,* 119.

[3] de Perrot M., Brundler M., Totsch M., Mentha G., Morel P.: "Mesenteric cysts, toward less confusion?". *Dig. Surg.*, 2000, *17,* 323.

[4] Bowen B., Ros P.R., McCarthy M.J., Olmstead W.W., Hjermstad B.M.: "Gastrointestinal teratomas: CT and US appearance with pathologic correlation". *Radiology*, 1987, *162*, 431.

[5] Prieto M.L., Casanova A., Delgado J., R. Zabalza: "Cystic teratoma of the mesentery". *Ped. Rad.,* 1989, *19*, 439.

[6] Dehner L.P: "Cystic teratomy of the mesentery". *Hum. Pathol.,* 1983, *14*, 493.

[7] James M.C.F. Jr.: "Dermoid cysts of the mesentery - Case report". *Am. J. Surg.,* 1944, *65*, 116.

[8] Buonanno G., Gonnella F., Pettinato G., Castaldo C.: "Autoimmune hemolytic anemia and dermoid cyst of mesentery. A case report". *Cancer,* 1984, *54*, 2533.

[9] Sharma V.P., Vyas K.C., Soni B.M., Purohit S.S.: "Mesenteric cysts". *J. Indian Med. Assoc.,* 1986, *84,* 185.

[10] Walker A.R., Putman T.C.: "Omental, mesenteric and retroperitoneal cysts - a clinical study of 33 new cases". *Ann. Surg.,* 1973, *178*, 13.

[11] Chiba T., Iwami D., Kikuchi Y.: "Mesenteric teratoma in an 8-month old girl". *J. Ped. Surg.,* 1995, *30,* 120.

[12] Verswijvel G., Janssens F., Vanboven H., Palmers Y.: "Spontaneus rupture of mesenteric dermoid cyst - a rare cause of abdominal pain". *Eur. Radiol.,* 2004, *14,* 1517.

[13] Hastings N., Norris W.J.: "Unusual abdominal cysts in infants and children". *Cal Med.,* 1954, *81,* 84.

[14] Outwater E.K., Siegelman E.S., Hunt J.L.: "Ovarian teratomas-tumor types and imaging characteristics". *Radiographics,* 2001, *21*, 475.

[15] Bruneton J.N., Benattar E., Drouillard J., Balu-Maestro C., Normand F.: "Mesenteric cysts in the adult". *J. Radiol.,* 1987, *68,* 687.

[16] Lazar C., Chifan M., Strat V., Diaconescu M.R., Tircoveanu E., Luchian S., Dobrescu G.: "Tumors and cysts of the mesentery. Clinical and therapeutic study". *Rev. Chir. Oncol. Radiol.*, 1982, *31,* 423.

[17] Parlakgumus A., Tokmak M., Bal N., Ezer A., Yildirim S.: "Diagnosis and treatment of mesenteric tumors". *Acta Chir. Belg.*, 2010, *110,* 51.

[18] Shin N.Y., Kim M.J., Chung J.J., Chung Y.E., Choi J.Y., Park Y.N.: "The deferential imaging features of fat containing tumors in the peritoneal cavity and retroperitoneum - the radiologic-pathologic correlation". *Korean J. Radiol.,* 2010, *11, 3*33.

[19] Wang P.C., Yang T.L., Pan H.B.: "Ct images of a malignant transformed ovarian mature cystic teratoma with rupture: a case report". *Korean J. Radiol.*, 2008, *9,* 458.

[20] Yoo E., Kim M.J., Kim K.W., Chung J.J., Kim S.H., Choi J.Y.: "A case of mesenteric cystic lymphangioma - Fat saturation and chemical shift MR imaging". *J. Magn. Reson. Imaging,* 2006, *23*, 77.

[21] Okamoto D., Ishigami K., Yoshimitsu K., Irie H., Tajima T., Nishic A. *et al.*: "Hemorragic mesenteric cystic lymphangioma presenting with acute lower abdominal pain: the diagnostic clues on MR imaging". *Emerg. Radiol.*, 2009, *16*, 327.

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