Intra-abdominal lymphatic malformation

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Abstract

Intra-abdominal lymphangioma is a rare congenital lymphatic vascular malformation. It is found, most commonly, in adult patients who present with abdominal pain, and whom medical imaging (ultrasound, endoscopy and tomography) reveals a cystic intra-abdominal mass. Such masses may be on the mesentery, in contact with abdominal viscera or may be part of the visceral wall. Diagnosis of intra-abdominal lymphangioma is difficult because of its rarity, and because other pathologies may have the same radiological signs. The diagnosis is confirmed by anatomopathological examination with immunohistochemistry. The treatment is complete surgical excision, in order to prevent recurrence. (Acta gastroenterol. belg., **2006**, 69, **209-212**).

Key words : cystic lymphangioma, vascular malformation, intra-abdominal.

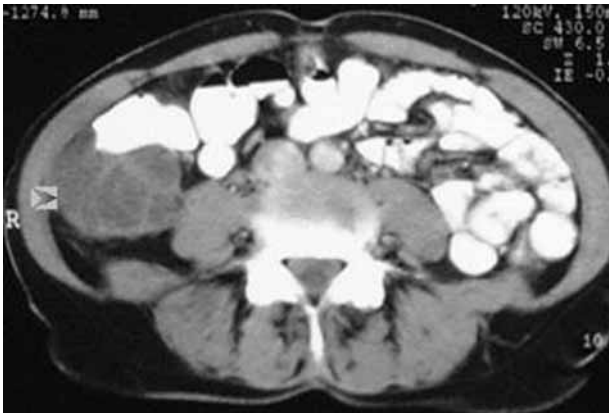

Fig. 1. — Abdominal Tomography, cystic mass marked by an arrow.

Introduction

The discovery of a cystic mass in the mesentery, or in contact with an intra-abdominal organ is not exceptional (1,2). The nature and classification of such masses remain poorly understood because of the low incidence of each anatomopathological type (1). The lack of recognition of their existence may lead to errors in their management. Of these cystic masses, lymphatic vascular malformation is particularly uncommon (3-17), and their classification has rapidly improved with electronic microscopy and the advent of immunochemical techniques.

Case report

A 54 years old lady was admitted to the emergency department, complaining of pain in the right iliac fossa, evolving for 3 days. She did not have nausea or vomiting. In her past medical history is abdominal surgery for a perforated ulcer. She had no known drug allergies and was taking no medication. On physical examination, her temperature was 36.7°C, her blood pressure of 120 mm Hg systolic, 80 mm Hg diastolic. Heart rate is of 80 cpm. The patient was not cyanosed, was breathing normally, and well-hydrated. Cardiopulmonary examination was normal. Abdomen examination revealed normal peristalsis on auscultation. On palpation the patient had a soft belly with a tender area in the right iliac fossa, a little higher and lateral to the Mac Burney's point, with a significant and mobile mass.

Haematological investigations showed 9.200 leucocytes/mL, 68% neutrophils, C-Reactive Protein 3.39 mg/dL, D-dimers 2616 ng/mL. Blood electrolytes were 142 mEq/L sodium, 3.21 mEq/L potassium, 92mEq/mL chlorine, other biological parameters were normal. Urine analysis showed white blood cells 3 crosses, red blood cells 3 crosses, protein 2 crosses and glucose 1 cross, culture was negative.

Abdominal ultrasound was performed, and showed a liquid formation, with intralesional strands, which measured approximately 9.6×4.0 cm in the right iliac fossa, in contact with the cæcum and the lower pole of the right kidney, the respect of the liver, the gall bladder, and the spleen. Small cortical cysts were present on both kidneys. An abdominal Tomography without and with injection of contrast, and opacification of the digestive loop by diluted contrast medium (Fig. 1). It shows a regular liquid formation presenting intra-lesional strands, in the region of the right iliac fossa and extending from the lower pole of the right kidney down to the appendix. Tomography confirmed small cortical cyst on both kidneys, and the respect of the liver, the gall bladder, the

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Submission date : 02/04/2005.
Acceptance date : 08/11/2005.

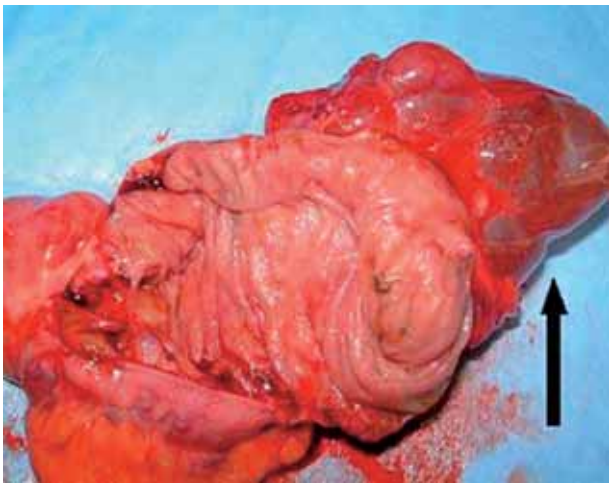

Fig. 2. — Surgical resection of colon cystic masses marked by an arrow.

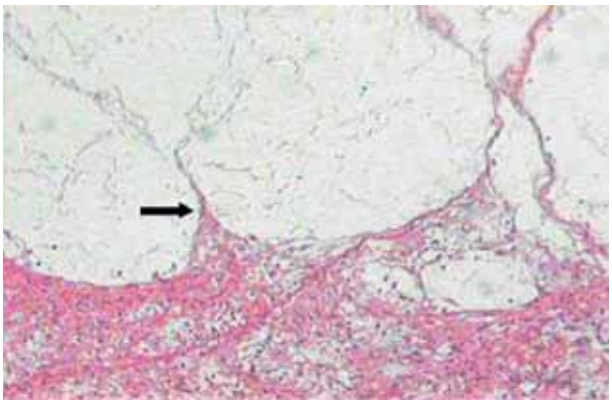

Fig. 3. — Histopathological findings : Cysts (Hematoxylin and eosin coloration), the endothelial layer marked by an arrow.

pancreas, supra renal glands, the spleen and annex bodies ; presence of a liquid blade on the Douglas. These two radiological examinations show a probable appendicular mucocoele or even a peritoneal pseudomyxome.

The patient was hospitalized and treated with low molecular weight heparin at a prophylactic dose and ranitidine 300 mg for gastric acid suppression.

In the light of raised D-dimer, a phlebography was carried out to exclude a thromboembolic phenomenon and was negative.

An exploratory laparoscopy was performed and confirmed the presence of the mass in the right parietocolic gutter impinging upon the ascending colon, to which it was intimately adhered. One of the cysts was haemorrhagic. A straight lines colectomy is carried out (Fig. 2), and the abdominal cavity was washed with an isobetadine solution.

The pathological examination shows a cystic lymphangioma with source of acute inflammation and haemorrhage. Post-operative recovery was completely satisfactory and the patient returned home on the 7th day post surgery.

Discussion

Until now lymphatic vascular malformations as lymphangioma were considered to be rare benign congenital tumors (2,3,7,8,11-13,17-26) due to sequestration of mesodermal tissue whose lymphatic connection to the systemic lymphatic network was failed (4,6,11,13-20,22). These lesions have been known for a long time as cystic lymphangioma. In the last 100 years several authors (e.g. Wegener (20,27,28), Redenmacker (14,29), Chisholm (5,10,16,20,23,24), Kaufman (16), Pean (6)) have described lesion of this type, at different places. The incidence (1/100.000) has increased (3,14,23,24), in line with the progress in diagnostic imaging techniques : ultrasound, endoscopy, magnetic resonance and tomography (5,16,17,22,24,26). In 1130 autopsies, Hora *et al.* found 6 abdominal lymphangiomas. This type of lesion is particularly well described in Japanese medical literature, where the incidence seems to be higher than in America or in Europe (5,9,10,17,20,22,24).

Several classifications exist, of which the best known distinguishes the capillary, cavernous and cystic lymphangiomas is from Wegener (20,27,28,30). In 148 abdominal lymphangiomas, Matsuda and al. count 104 cystic lymphangiomas, 43 cavernous and 1 simple lymphangioma (24).

These lesions can occur anywhere where lymphatic vasculature is located. The best known sites are the most accessible : the head, neck and in the axillae and lymphangiomas in this regions are usually diagnosed and treated in early childhood (2-7,11-16,19,21,29,31). The internal localizations, digestive, bone, or thoracic (32) are less common and tend to present in adulthood (7,8, 11,12,19,24,34). This type of congenital lesion can be diagnosed on prenatal ultrasound (2,4,19). Takuya Hatada and al, in a series of 128 cases discovered an average age for diagnosis of 51.2 ± 13.7 years (5,24), and with a sex ratio of 90 men for 33 women as Rieker *et al.* (14). Within the abdomen the cysts tend to be distributed in the mesentery (1,4,5,8,19,23,35), the small intestine (20,22,28), the colon (22,24), the rectum (20), the retro-peritoneum1 (3,22,30), the liver (15), the pancreas (15), the spleen (15), the appendix (15), the gall bladder (15), the supra renal glands (15), the duodenum (28) and the stomach (12). Of the colonic cysts, (Takuya Hatada and al.) 36% occur in the transverse colon, 29% ascending colon, 13% cæcum, 11% descending colon, 7% sigmoid colon, 4% rectum (5). Occasionally lymphangiomas are multiple, affecting several organs and is known as lymphangiomatosis (7,13,14,21,23,32,33). This condition is difficult to operate upon, and prognosis is poor (7,21,23,33).

Table I. — **Differential diagnosis of mesenteric cystic masses**

Diagnosis	Histological Aspect	Markers	Electronic microscopic aspect
Lymphatic Cyst	Flat endothelial cell	FVIII, CD31,CD34	Pinocytototic vesicles, tonofilaments and tight junction
Simple lymphatic cyst	„	„	„
Lymphangioma	„	„	„
Mesothelial cyst	Cubical cell type mesothelium	Cytokeratin, Vimentin	Surface border by microvilli and desmosomes between cells
Mesothelial simple cyst	„	„	„
Benign cystic Mesothelioma	„	„	„
Malignant cystic Mesothelioma	„ innumerable mitosis	„	„
Enteric cyst	–	–	–
Enteric duplication cyst	Enteric epithelium surface with smooth muscularis layer and neural complex	–	–
Enteric cyst	Enteric epithelium	–	–
Urogenital cyst	–	–	–
Mature Teratoma (dermoid cyst)	–	–	–
Pseudocyst non pancreatic	–	–	–
Cystic Tumours of pancreas	–	–	–

At the macroscopic anatomopathological level, the lymphatic malformations are bulky masses. Multicystic masses are more common than monocystic (1,7,8,17,20,23,30,35) and containing fluid who can be chylous if lesions sit at the proximal part of small intestine or near thoracic duct, at the other places it's a serous clear fluid (2,6,7,17,30). At the microscopic level (Fig. 3), one can observe several cavities composed of smooth muscle cells and collagen, and covered by endothelial cells with lymphocytes and *foam cells.*

Immunohistochemistry reveals the cells to be true endothelium, shown by endothelial tissue markers factor VIII , CD34 and CD31 antigens (8,25,33) and not a mesothelium which would be shown by marker for vimentin and cytokeratin (24,25). Electronic microscopy can distinguish cystic mesothelioma from cystic lymphangioma (25). In the first cells are border by a lot of microvilli, and intracytoplasmic tonofilaments in contact with desmosomes. In the second, cytoplasmic membrane is smooth, with pinocytotic vesicles (23), and intracytoplasmic fibrils and tight junctions (11,23). Christina Mahle *et al.* demonstrate that the proliferative activity is able to explain the growth of the cyst (8,21). Some authors have described malignant transformation of lymphangioma into lymphangiosarcoma after radiotherapy (14). A significant characteristic of this lesion in the abdomen is that they can occur just as much in the retroperitoneum (13), on the mesentery (25), on the serous surface of the viscera, as in the submucosa (5,8,9,10,12,15,16,17,24), where the lesion takes a intraluminal round aspect, smooth, translucent appearance with a broad base (17,22,24), as shown in some publications where the diagnosis was made by endoscopic resection (5,8,9,10,16,17,20,24,28,29).

The gastrointestinal symptomatology of these lesions is mainly due to their size expansion and their site (1,2,3,4,6,11,12,14,15). Complication may arise, such as infection (4,6,11,13,14,18,19,23,33), twisting (6,18), intussusceptions (5,9,10,17,20,22,24,29,32), stenosis (5), rupture (19,32), intra-cystic haemorrhage (1,3,6,7,31), hemoperitoneum (5), and hemoretroperitoneum (37). Melena (5,16,22,24,28) can occur when the lesion take place in the submucosa, like digestive enteropathy (6,7,13,14,15,16,1718,22,24). The symptoms may then be abdominal pain, or simple digestive trouble to acute abdominal pain (2-9,11,12,14,17, 18,19,20,22,28,31,35,36). The cardinal symptoms are pain with a palpable mass (2,3,6,7,11,12,15,17,19,22, 32,36), associated with nausea and vomiting (36).

The differential diagnosis of these mesenteric cystic masses was particularly well described by Perrot and Ros (1,25) (Table 1).

The best diagnostic techniques are ultrasound, CT (13) and MR (1,4,6) which is particularly useful for the detection of intra-cystic haemorrhage (3,6). They can be detected as hypersignal in T1-and T2-weighted sequences, while a serous content is hyposignal in T1-weighted sequences and hypersignal in T2-weighted sequences (7). For intraluminal lesions, barium enema with double contrast is possible but has been largely replaced by flexible endoscopy (5,9,17,22,24,26).

The diagnosis is confirmed only by pathology.

In terms of treatment, the major difficulty has been to differentiate between benign and malignant types

(25,38) (mesothelioma or sarcoma), for which the treatment differs (1,3,18). It is widely agreed that complete excision is required to avoid recurrence or complications (3,4,6,8,11,12-14,17-19,23,30,31,35,36), and sometimes adjacent structures must also be removed1 in order to ensure this, but vital organ must be share (3,6,20,25,35). The surgical method is subject to debate : some authors advocate resection by endoscopy for intraluminal lesion less than 2 centimeters (5,9,10, 16,17,22,24,26), while others advice the sclerotherapy technique (5,6,19,24,30). The diffuse nature of the lesions, the risk of recurrence (2) and uncertainty over the exact type of tumours necessitates caution, lead us towards complete, and intact excision (3,6,20), and only in case of inextirpable lesion, the sclerotherapy may be an indication (4,5,19).

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