A rare case of giant mesenteric cystic lymphangioma of the small bowel in an adult: A case presentation and literature review

Z. Mehmedovic[1], M. Mehmedovic[2], M. Konrad Custovic[3], A. Sadikovic[3], N. Mekic[4]

(1) Department of General Abdominal Surgery, Clinic for Surgery, University Clinical Center Tuzla, Tuzla, Bosnia and Herzegovina ; (2) Department of Gastroenterology and Hepatology, Clinic for Internal Diseases, University Clinical Center Tuzla, Tuzla, Bosnia and Herzegovina ; (3) Department of Pathology, Polyclinic for Laboratory Diagnostics, University Clinical Center Tuzla, Tuzla, Bosnia and Herzegovina ; (4) Primary Healthcare Center "Dr. Mustafa Sehovic" Tuzla, Tuzla, Bosnia and Herzegovina

Abstract

Background : **Lymphangiomas are tumours that are often found in the head and neck in children and, rarely, in adults. Among all lymphangiomas in adults, the small bowel mesentery type accounts for less than 1%. We aim to present a rare case of small bowel symptomatic cystic lymphangioma in an adult patient, as well as its diagnosis and subsequent treatment.**

Patient : **An adult female who was admitted to the University Clinical Center Tuzla for surgery presented with abdominal pain, tenderness and swelling of the abdomen. A giant cystic small bowel mesentery lymphangioma, which was histopathologically confirmed, was enucleated during open surgery.**

Conclusion : **Timely diagnosis and treatment of this rare condition are crucial. Although benign in nature, mesenteric lymphangiomas may cause significant morbidity or mortality due to their huge size and critical locations.** (Acta gastroenterol. belg., **2016**, 79, **491-493**).

Key words : cystic lymphangioma, mesentery, small bowel.

Introduction

Cystic lymphangiomas are uncommon mesenteric tumours. They are preferentially located in the head, neck and axilla in children (1, 2, 3). Lymphangiomas in the peritoneal cavity are extremely rare, especially in adults (2, 4), while small bowel mesentery lymphangiomas account for less than 1% of all lymphangiomas (2, 3). Intra-abdominal mesenteric lymphangiomas have an incidence of approximately 1/20 000 to 1/250 000 hospital admissions (5). Intra-abdominal lymphangioma usually presents as abdominal distension, a palpable abdominal mass or acute intestinal obstruction (3)**.** An abdominal ultrasonography (US), a computed tomography (CT) scan and a celioscopy might be useful for establishing the diagnosis (1, 6), whilst the treatment is mainly surgical (7). We report a case of an adult Caucasian female who was admitted to the clinic for surgery, with clinical signs of a palpable abdominal mass, which was later confirmed as a giant cystic lymphangioma.

Case report

A Caucasian female, who was 35 years of age, was referred to the clinic for surgery because of a dull abdominal pain and swelling of the abdomen in the region of the epigastrium. She was showing signs of a palpable abdominal mass. Her previous medical history revealed two caesarean sections without any other diseases to report. Laboratory tests were performed and all of the parameters were within their normal range of values (complete blood count, blood glucose, parameters of renal function and C-reactive protein). The tumour markers (alpha-fetoprotein (AFP), carcinoembryonic antigen (CEA), carbohydrate antigens CA 19-9, CA 125, and CA 15-3) were negative. After performing an ultrasound examination, a multicystic formation was identified, which was 17cm in diameter. A CT-scan was performed, which identified the lobular cystic formation of fluid density, 17x8x17cm in size (Figure 1). During surgery, a small bowel mesentery cystic formation of the same size was found (Figure 2A). This was filled with a milk-like fluid. An enucleation of the formation was performed, leaving the surrounding anatomical structures intact. No complications in postoperative recovery were noticed. An histopathology analysis confirmed mesenteric cystic lymphangioma (Figure 2B)**.** An immunohistochemical analysis for endothelial markers CD31 and CD34 showed diffused positivity in the cystic wall endothelium (Figure 2C). The lining cells of the cystic walls were non-immunoreactive for calretinin. The patient has undergone regular check-ups without any signs of a recurrence of the tumour.

Discussion

Intra-abdominal mesenteric lymphangiomas are uncommon tumours, which usually manifest in early adulthood. Although intra-abdominal cystic lymphangiomas in adults are very rare, reviewed literature on this matter shows that, in the last decade, there has been an increase in reporting of the condition. This case report sums up the main features and conclusions from literature that have been published thus far. As

Correspondence to :Zlatan Mehmedovic, M.D., University Clinical Center Tuzla. Clinic for Surgery, Department of General Abdominal Surgery. Trnovac bb, Tuzla 75 000, Bosnia and Herzegovina.
E-mail: zmehmedovic@hotmail.com

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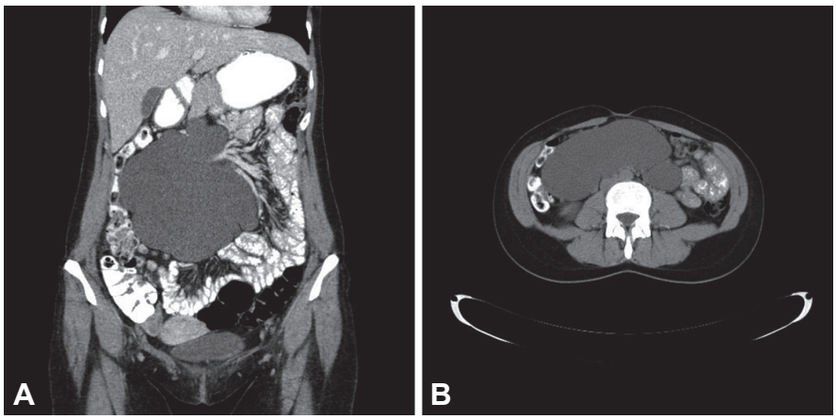

Fig. 1. — Native and contrast-enchanted scans of the abdomen show an oval fluid density formation, measuring 17x8x17cm of slightly lobular outlines. This extends from the caudal stomach outline to the upper pelvic aperture. By its bulkiness, this formation dislocates the leaning abdominal structure – stomach, small and large intestines, and mesenterial vascular structures. Coronal (A) and axial (B) planes.

mentioned, small-bowel mesentery lymphangiomas are responsible for less than 1% of all lymphangiomas (3). Three forms of lymphangiomas are described as: (i) Capillary lymphangioma, (ii) Cavernous lymphangioma and (iii) Cystic lymphangioma (2, 8). In our case, histopathology confirmed cystic lymphangioma. The cystic type consists of lymphatic spaces of various sizes, which contain fascicles of smooth muscle and collagen bundles, but has no connection with the adjacent normal lymphatics. Cystic lymphangioma is not always clearly differentiated from the cavernous type, as the cystic type may also contain cavernous areas (2).

A well-established theory suggests that lymphangiomas arise from sequestrations of lymphatic tissue during embryologic development. It is also suggested that abdominal trauma, lymphatic obstruction, inflammatory processes, surgery or radiation therapy may lead to the secondary formation of such a tumour (4). The cystic lymphangioma was not identified at the time of the two previous pregnancies, which were both performed via caesarean sections. Therefore, this cystic mass could be a consequence of the previous abdominal surgery.

Cystic lymphangiomas may manifest via acute abdominal pain. This is associated with bowel obstruction, signs of peritonitis and a chronic abdominal swelling, which is detected by a palpation of a cystic mass or abdominal swollenness with lower extremities lymphoedema (6).

The differential diagnosis of an intra-abdominal cystic mass includes an enteric duplication cyst, an enteric cyst, a mesothelial cyst, a pancreatic pseudocyst, a non-pancreatic pseudocyst, a cystic mesothelioma, a cystic spindle cell tumour and a cystic teratoma (9).

An abdominal US, CT-scan and celioscopy might be useful for establishing the diagnosis (1). In our case, an abdominal US and CT were sufficient to elect surgical treatment. Although US and CT are the most commonly used diagnostic tools, preoperative diagnosis is only possible in 22,6% of cases (10).

The US method shows mesenteric lymphangioma as a cystic lesion with multiple thin septae. Meanwhile, CT imaging projects a uni- or multilocular mass with enhancement of the wall and septum by contrast medium (2).

Although rare in incidence, these lesions have been shown to be accurately diagnosed with an endoscopic ultrasound guided fine-needle aspiration (EUS-FNA). Aspiration draws out a milky-white fluid, which is notable for lymphocytes with fluid trygliceride levels that are elevated above 500 mg/dl (11).

Although imaging methods are a useful diagnostic tool, definitive diagnosis is confirmed by histopathology after a complete surgical resection (5).

The diagnosis of cystic lymphangioma is confirmed using three standard histological criteria: (i) cyst lined by a flat endothelial epithelium, (ii) small lymphatic spaces and (iii) abundant lymphoid tissue (12).

An immunohistochemical analysis for endothelial markers CD31 and CD34 showed diffused positivity in the cystic wall endothelium, supporting the diagnosis of our patient. Lymphoid follicles and lymphoid infiltrates in the stroma, also supported the diagnosis. The lining cells of the cystic walls were non-immunoreactive for calretinin. Therefore, benign multicystic mesothelioma as a differential diagnosis was excluded (3, 6, 13).

The primary treatment of mesenteric cysts is a surgical resection. Traditionally, this has been through open surgery. However, laparascopic resection has also been reported (7). When feasible, surgery consists of an enucleation. The segmental intestinal resection is achieved when the cyst adheres intimately to the bowel (1, 13).

Recent literature has cited alternatives to surgery for the unresectable disease. These include the percutaneous

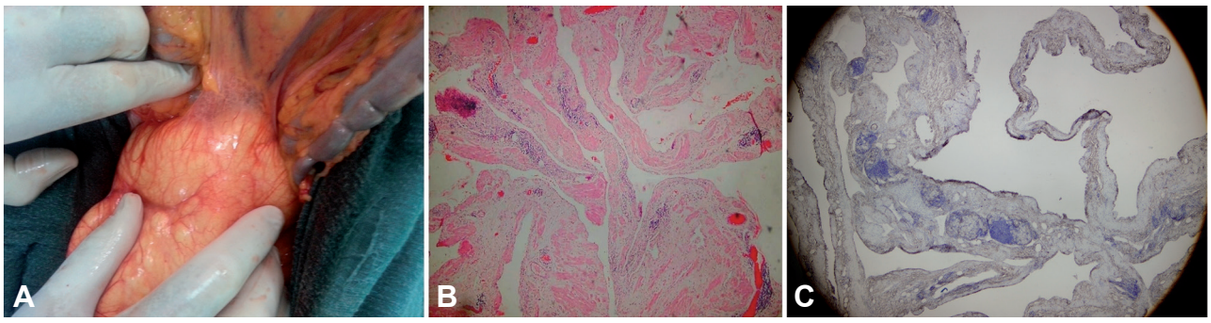

Fig. 2. — Intraoperative image shows cystic tumour formation of the small bowel mesenterium, measuring 17cm in diameter during its enucleation (A). The received specimen (H&E, 4x) partly consists of adipose tissue and partly of dilated thin-walled spaces, which are filled with eosinophilic fluid and lined by flat endothelial cells. The intervening stroma contains scattered lymphoid aggregates and wisps of smooth muscle fibres (B). An immunohistochemical analysis for CD34 showed diffused positivity in the cystic wall endothelium (C)

intracystic injection of agents such as alcohol (12). OK-432 is a sclerosing agent that consists of lyophilized streptococcal preparation. This induces a strong local inflammatory reaction. This is used mainly for neck cystic lesions (14).

Local recurrence of the tumour is possible (6). Our patient was re-examined a year after surgery and no recurrence of the tumour was identified.

Conclusion: Mesenteric cystic lymphangioma is very rare in adults. Timely diagnosis, as well as treatment of this rare condition, is critical in preventing significant morbidity or mortality, which is caused by the size of the cyst and unfavourable locations.

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