Mesenteric cystic lymphangioma causing intestinal occlusion in an adult patient

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Summary: Mesenteric cystic lymphangioma causing intestinal occlusion in an adult patient.

Introduction

Cystic lymphangiomas are very rare benign tumors of vascular origin. In 90% of cases, they occur in children aged < 2 years (8), while in adult population they have an incidence of 0.4-1 cases every 100,000 hospitalizations (7). Most frequent localizations are the neck (70-75%) and the axillary region (20%); in 5% of cases they can be detected in the retroperitoneum, mesentery, mediastinum, lung, limbs, thoracic wall, lumbar spine, liver, spleen, pancreas, and in the inguinal region (2, 4, 9, 11, 13).

Lymphangiomas are to be considered benign tumors of the lymphatic system whose etiology is still unclear. According to more recognized dysembryogenetic theories, the origin of this pathology is attributed to an error of the sectorial development of the lymphatic system whose gems, constituting the lymphatic capillary network of an organ, become independent, thus proliferating and forming multiple cysts which lose the connection between the lymphatic and the venous system (1, 3, 11).

Other theories consider the onset of lymphangiomas following traumas (10), inflammatory processes, or localized lymphatic degenerations (9).

Histologically, lymphangiomas can be divided into three types: lymphangioma circumscriptum, characterized by multiple clusters of vesicles; cavernous lymphangioma, which can become huge and can recur very easily if not completely excised; cystic lymphangioma, a multilocular tumor, which grows very slowly but can reach very large size, usually clearly-defined and well-removable (3) but which can recur if not radically excised.

Caso clinico

L.I., a 46-year-old man, had already undergone an urgent subtotal colectomy with resection of the bladder dome eight years earlier for intestinal occlusion due to a carcinoma of the sigmoid colon (pathologic classification: pT3pN0pMx). Surgery was followed by a
Discussion

Because of its rare incidence and its onset nearly exclusively in pediatric age (90% of cases) (5, 6), cystic lymphangioma of the mesentery is included in the differential diagnosis of endoluminal cysts in adult, also because an abdominal localization occurs in 2-8% of cases only. Cystic lymphangiomas are asymptomatic long time before displaying rather aspecific symptoms characteristic by intestinal sub-occlusion or, more rarely, by acute abdomen and acute intestinal occlusion due to intra-peritoneal rupture of cyst, intracystic bleeding, infection, or intestinal volvulus (5).

The pre-operative diagnosis is troublesome and it is based on investigations such ultrasound, CT-scan and MR of the abdomen, able to reveal retroperitoneal localization in the mesenteric root and the cystic nature of the lesion, which has multiple septa and liquid content. A differential diagnosis of cystic lymphangiomas should include: hydatid cysts, cysts and pseudocysts originating from contiguous organs, pseudocysts due to blood collections, and malformative cysts, such as dermoid cysts, enterocystomas, cysts arising from remnants of the wolffian duct, and urachal cysts. In nearly all cases, however, a definitive diagnosis is provided by the histology of the surgical specimen.

The treatment is exclusively surgical, with the complete excision of the cystic wall to avoid any recurrence. Because of the benign nature of the neof ormation, there is no trend to infiltrate the surrounding structures and, in most cases, the lesion is completely removable (12). In case of involvement of mesenteric vascular structures and ileal loops, it might be necessary to proceed to the resection of a loop with immediate restoration of the intestinal continuity. In other few cases, as in the patient presented here, because of the extensive involvement of the mesenteric root, the radical surgical excision is not possible. Furthermore, the effectiveness of radioderapy or chemoterapy for this pathology has not been proven.

In our case, a pre-operative diagnosis was not possible and all the symptoms reported by the patient were mostly attributed to the previous surgery, while all pre-operative investigations performed until five days before the intervention did not show the cystic nature of the lesion, which was at first believed to an abnormal distension of the pre-anastomotic ileal loop; furthermore, the surgical resection of the lesion was not radical because of the adhesion with surrounding vascular structures and ileal loops.

A careful and intensive follow-up by US, CT, and MR is required to evaluate the clinical course of the cystic lymphangioma.
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Bibliografia