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casistica clinica

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.**

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Cystic lymphangioma is a benign tumor of uncertain etiology characterized by a slow growth; in 2-8% of cases it is localized in the mesentery. Symptomatology is aspecific and preoperative diagnosis is often difficult.

The Authors report the case of a mesenteric cystic lymphangioma in a patient who had undergone subtotal colectomy eigh years earlier for an adenocarcinoma occluding the sigmoid colon. The patient was hospitalized for intestinal occlusion.

RIASSUNTO: **Il linfangioma cistico del mesentere quale causa di occlusione intestinale in un paziente adulto.**

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Il linfangioma cistico è un tumore benigno ad etiologia ignota, caratterizzato da lenta crescita; solo nel 2-8% dei casi si localizza a livello del mesentere. La sintomatologia è aspecifica e la diagnosi preoperatoria è spesso difficile.

Gli Autori riportano il caso di un linfangioma cistico del mesentere in un paziente già sottoposto a colectomia subtotale otto anni prima per un adenocarcinoma occludente il sigma. Il paziente fu ricoverato per una occlusione intestinale.

KEY WORDS: Cystic lymphangioma - Intestinal occlusion - Mesentery root.
Linfangioma cistico - Occlusione intestinale - Mesentere.

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 dysembryiogenic 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 loose the connection between the hymphatic 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

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six-months chemotherapic treatment. Periodical total body CT exams, rectoscopies, and tumoral markers never showed any recurrencies, but only the distension of the pre-anastomotic tract. In the immediate post-operative period, the patient complained of sub-occlusive episodes alternating with diarrhoic discharges, attributed to adhesions. At clinical examination the patient reported weight loss, rectal bleeding, dizziness, and back pain since eight months. Total body CT-scan, MR of the spine, and rectoscopy were negative.

Because of the onset of ingravescent abdominal pain and total intestinal occlusion, the patient was hospitalized in our Department. He had a suffering aspect, a distended abdomen with diffuse abdominal pain and absence of peristalsis.

Laboratory findings revealed moderate anemia, neutrophilic leucocytosis, and low levels of potassium, while other imaging investigations, such as the x-Ray of the abdomen and the abdominal CT-scan showed high air-fluid levels in the mesogastric region. These were believed to be a huge distension of the pre-anastomotic ileal loop (Fig. 1). After 24 hours of infusional therapy, the symptomatology had improved and the patient was discharged. Four days later, however, because of the worsening of abdominal pain, the persistence of intestinal subocclusion and the worsening of general conditions, the patient was once again hospitalized in our Department. He presented wih fever, tachypnea and remarkable leucocytosis; a repeated x-Ray of the abdomen showed increased air-fluid levels as compared with the previous exam and a elevation of the dome of the diaphragm.

The patient, therefore, underwent an urgent explorative laparotomy. The incision of peritoneum, with difficult lysis of adhesions, revealed a grouping of ileal loops under which a cystic neoformation, measuring 25 cm in diameter and located at the mesenteric root, was present. This did not have a cleavage plane with the surrounding structures, especially the vascular ones (aorta and inferior vena cava), and its spontaneous rupture produced the leaking of a great amount of a liquid and revealed the presence of internal septa. Because of the impossibility of the complete excision of the cystic lesion, this was opened and drained externally. The bioptic exam of the surgical sample confirmed the diagnosis of infected cystic lymphangioma.

The post-operative course was uneventful. On the third day after surgery, the drainage of the cystic cavity was removed; it drained only a small amount of blood serum. The patient was discharged eight days after and, two years later only he is in good general conditions, although still complaining of episodes of abdominal pains and sub-occlusive disorders.

A CT-scan annd a MR of the abdomen carried out approximately one year after surgery did not reveal the presence of endoabdominal and/or retroperitoneal lesions.

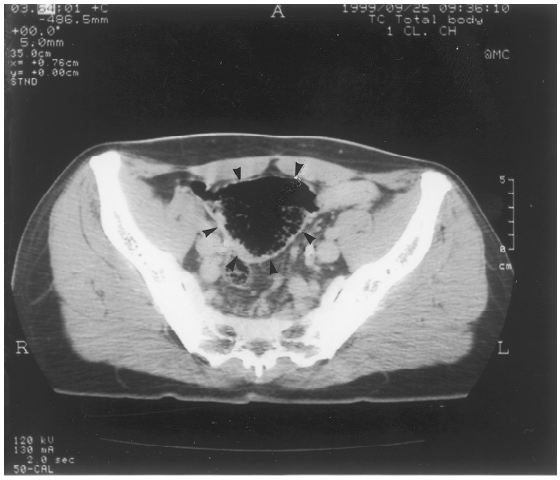

Fig. 1 - CT-scan: cystic lymphangioma of the mesentery.

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 rarely included in the differential diagnosis of endoabdominal 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 characterized by intestinal sub-occlusion or, more rarely, by acute abdomen and acute intestinal occlusion due to intra-peritoneal rupture of the cyst, intracystic bleeding, infection, or intestinal volvulus (5).

The preoperative diagnosis is troublesome and it is based on investigations such ultrasound, CT-scan and MR of the abdomen, able to releave 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 neoformation, 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 radioterapy 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 the 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.

Bibliografia

1. Barnett LA: Retroperitoneal cystic lymphangioma. JAMA 1960; 173:1111-1116.
2. Colizza S, Tiso B, Bracci F, Cuderno RG, Rigotti A, Crisi E: Lymphangioma of the stomach and jejunum: report of one case. J Surg 1981; 17: 169-176.
3. Colombo C, Paletto AE, Maggi G, Masenti E, Massaioli N: Trattato di chirurgia. Minerva Med 1993; 1: 240, 626.
4. Drago JR, De Muth WF Jr: Lymphangioma of the stomach in a child. Am J Surg 1979; 131: 605-606.
5. Fundarò S, Medici L, Perrone S, Natalizi G: Linfangioma cistico del mesentere. Minerva Chir 1998; 53: 939-942.
6. Gar-Yang Chau, Kwang-Lian King, Cheng-Hsi SU, Wing Yui Lui: Retroperitoneal cystic lymphangioma in adults. Int Surg 1993; 78: 243-246.
7. Kurtz RJ, Heimann TM, Bech AR, Holt J: Mesenteric and retroperitoneal cysts. Ann Surg 1986; 203: 109-112.
8. Rekhi BM, esselstyn CB Jr, Levi I: Retroperitoneal cystic lymphangioma: report of two cases and review of literature. Cleve. Clin Q 1972; 39: 125-128.
9. Roismann I, Manny J, Fields S: Shiloni. Intra-abdominal lymphangioma. Br J Surg 1989; 76: 485-489.
10. Srano RC, Carter BL, Bankoff MS: Cystic lymphangiomas: CT diagnosis. Br J Radiol 1984; 57: 424-426.
11. Smaili M, Ollivry J, Empinet O, Bouvier S, Lehur P, Le Borgne A: Les lymphangiomes kystiques du colon. J Chir Paris 1996; 133: 12-126.
12. Takiff H, Calabria R, Yin L, Stabile BE: Mesenteric cysts and intra-abdominal cystic lymphangiomas. Arch Surg 1985; 120 (11): 1266.
13. Vezzoi M, Ottini E, Montagna M, La Fianza A, Palli M, Rosso R, Mazzone A: Limphangioma of the spleen in an elderly patient. Haematologica 2000; 85 (3): 314-317.