CASE REPORT

Lymphangioma of the small bowel mesentery in a 7-year-old girl: a report of a case investigated with ultrasonography, elastosonography and computed tomography

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Abstract

Purpose To describe the characteristics of lymphangioma reporting a pediatric case of small bowel mesentery location, showing ultrasound-elastosonographic and computed tomography findings.

Methods A 7-year-old girl suffering from vomiting and abdominal pain underwent chest–abdomen radiography and abdomen ultrasound examination at our institution. These exams were repeated for 5 days after medical therapy and a computed tomography investigation was also performed.

Results Abdominal ultrasound and computed tomography examinations revealed the presence of a heterogeneous mesenteric mass occupying the right hemiabdomen displacing small bowel loops. Moreover, the mass had an irregular vascularisation pattern on color-Doppler study and appeared mostly green colored, like soft tissues, on elastosonographic colorimetric map. The little patient underwent surgery with removal of both the mass and the close adherent ileal loop. Finally, anatomopathologic diagnosis of mesenteric lymphangioma was made.

Conclusions Although mesenteric lymphangioma is a rare disease that presents neither clinical nor imaging typical features requiring invasive procedure to be diagnosed, ultrasound-elastosonographic findings can suggest a

benign condition and computed tomography scan is useful in order to plan surgery.

Keywords Lymphangioma · Elastosonography · Small bowel mesentery

Riassunto

Scopo descrivere le caratteristiche del linfangioma riportando un caso pediatrico di tumore localizzato nel mesentere del piccolo intestino, descrivendo i reperti alla tomografia computerizzata ed all'ecografia-elastosonografia.

Metodi una bambina di 7 anni affetta da vomito e dolore addominale è stata sottoposta a radiografia del torace - addome ed ecografia addominale presso il nostro istituto. Questi esami sono stati ripetuti cinque giorni dopo la terapia medica ed è stato anche eseguito un esame di tomografia computerizzata.

Risultati l'ecografia e la tomografia computerizzata dell'addome hanno rivelato la presenza di una massa mesenterica eterogenea che occupava l'emiaddome di destra spostando le anse del piccolo intestino. Inoltre, la massa presentava un pattern di vascolarizzazione irregolare allo studio color-Doppler ed appariva per lo più di colore verde, come i tessuti molli, alla valutazione elastosonografica. La piccola paziente è stata sottoposta ad intervento chirurgico con asportazione sia della massa che dell'ansa ileale aderente. È stata ottenuta, infine, la conferma anatomopatologica di linfangioma mesenterico.

Conclusioni sebbene il linfangioma mesenterico sia una malattia rara che non presenta caratteristiche tipiche né cliniche né di imaging, e che richieda un approccio invasivo per essere diagnosticata, l'ecografia-elastosonografia può suggerirne la natura benigna mentre la tomografia computerizzata si rivela utile al fine di pianificare l'intervento chirurgico.

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Introduction

Lymphangioma (LA) is a benign proliferation of thin-walled lymphatic tissue believed to originate from the early failure of communication of lymphatic spaces with the normal main draining lymphatic vessels, thus being considered a congenital malformation rather than an acquired tumor [4, 6, 7]. It is usually found in the head and neck (50–75 %) and axillary (20 %) regions during the first few years of life, while other locations are more rare [4, 7]. Among these ones, LA of small bowel mesentery represents less than 1 % of all cases described in literature [7]. The clinical presentation of abdominal LAs is polymorphic, varying from the incidental discovery of an abdominal cyst, which becomes symptomatic when reaches large size, up to the signs and symptoms of acute abdomen, with abdominal pain and bowel occlusion [3].

Case report

We report a case of a 7-year-old girl who was brought to the emergency room of our institution after 5 days of unexplained intermittent abdominal pain, vomiting, and irregular bowel without fever. The patient's vital signs were regular except for the pulse rate 120 beats/min, while physical examination revealed abdominal distension without rigidity or masses. Other features were unremarkable and blood tests were not altered. Plain chest-abdomen radiography (XR) and ultrasound examination (US) of the abdomen were performed. XR pathological findings were gaseous bowel distension mainly of the colon and minimal gas–fluid levels. US revealed a heterogeneous hyperechoic mass located in mesogastric–hypogastric regions behind and upper the bladder, characterized by several hypoechoic–anechoic areas and a disorganized vascular pattern on color-Doppler evaluation. The mass was initially believed to originate from the ovary and the child was hospitalized and posted to oncologic division of our institution. Her symptoms were temporarily relieved by treatment with painkilling and anti-vomiting agents. Five days later, abdomen XR and US were repeated. XR was negative for loop distension and levels and so US (Esaote-MyLab, linear probe 4–13 MHz) allowed to appreciate an irregular hyperechoic peritoneal sheet thickening associated to alternated hypo-anechoic lacunar areas, branching among intestinal loops in the right quadrants of the abdomen. The mass was far about 3 cm in depth from cutaneous plane, and had an irregular vascularisation pattern on color-Doppler study. The study was completed with visual elastosonographic evaluation of the mass which appeared mostly green with some red areas and transient blue spots on conventional color map of tissue stiffness orienting for a

prevalent soft consistency of the lesion (Fig. 1a). Several lymph nodes were found in the mesentery and all of them were not larger than 1 cm in maximum diameter. Although these findings suggested an intra-abdominal mass containing cystic areas, they did not allow us to have an univocal interpretation. Because a further investigation was required also to better evaluate the precise site and the extent of the lesion, a contrast enhanced computed tomography (CT) study was executed. It confirmed the presence of a mesenteric mass which appeared to originate from mesentery root between celiac trunk and the origin of the superior mesenteric artery and extended in the mesentery reaching the small bowel wall. The mass was heterogeneously hypodense with fatty and calcific components, and had no contrast enhancement, and occupied the right hemiabdomen displacing small bowel loops and caudally reached the bladder dome (Fig. 1b, c). US and CT findings seemed insufficient to provide a definite diagnosis requiring the removal and examination of a tissue sample. However, considering both clinical past events of the child and the extent of the mass in close contact with a bowel loop, surgeons believed surgery was the better choice of management in order to avoid obstructive complications like volvulus. Then, the child underwent laparotomy. The mass was found between the sheets of the mesentery appearing yellowish-white, hard and tenaciously adherent to the borders of the intestinal loops for 15 cm in length. The mesenteric mass with adjacent small bowel segment was resected and a ileo-ileal termino-terminal anastomosis was performed (Fig. 1d). Cut surfaces of the mass revealed multicystic spaces of varying size containing milky white fluid. Microscopic examination showed the presence of several dilated vascular channels of various diameters, defined by walls comprising smooth muscle and lined with flat endothelial cells. The stroma contained scattered lymphoid aggregates and reached the submucosa from serosal surface of the bowel. Also according to immunochemical analysis of CD31, CD34, SMA, Actina and CD45, finally the diagnosis was mesenteric LA.

Discussion

Lymphangioma is a mass-forming lesion which usually presents in the first few years of life with 50–60 % manifesting by age 1 year and 90 % by age 2 years, while in our case the little patient was 7-year-old [4, 6, 7]. Although the incidence of abdominal LAs is unknown, they represents for 3–9.2 % of all pediatric LAs, and among these, LA of the small bowel mesentery, has been described in less than 1 % of all LAs [3, 6, 7]. Histologically, LAs can be distinguished into three types: capillary or simple, cavernous, and cystic. Cystic spaces are seen for both cystic and

Fig. 1 Ultrasound examination of the abdomen (**a**): the mass appears heterogeneously hyperechoic with several hypo-anechoic areas (in the green square on the left) which are colored in red on elastosonographic map (on the right half of the image); computed tomography images: **b** the mass appears close to small bowel loops, extending

from superior mesenteric artery to the bladder dome, and **c** it is heterogeneously hypodense with fatty components; aspect of the lymphangioma after surgery (**d**): removed mass appears yellowish-white, tenaciously adherent to the borders of the intestinal loop for 15 cm in length

cavernous LA and may contain chylous, serous, hemorrhagic, or mixed fluid [3, 4]. In this case study, sectioned mass presented multicystic aspect with cysts containing milky white fluid supporting LA, in which the white color is due to the great presence of lymphoid cells. Microscopically, dilated lymphatic spaces of varying size were associated with smooth muscle cells and lymphoid aggregates clearly characteristic of cystic LA [4, 7]. Intra-abdominal LA usually presents with abdominal distension, a palpable abdominal mass, or acute intestinal obstruction when LA induces volvulus of the small bowel [3, 4, 7]. However, LA may not be palpable due to its flaccid and mobile nature [5]. In our case, child had vomiting and bowel distention but had neither a palpable abdominal

mass nor an intestinal obstruction but the fear of an obstructive evolution of mass growth influenced therapeutic decision. The diagnosis of mesenteric LA is usually made histologically following surgical resection of the mass. There are no blood tests which will establish the diagnosis. Radiological investigation usually confirms the presence of a mass and may help to exclude other causes of intra-abdominal masses but is usually insufficient to provide a definite diagnosis [1, 5, 7]. Plain XR may demonstrate a noncalcified soft-tissue mass or displaced bowel loops and possibly small bowel obstruction [2, 5]. The multilocular aspect of LA is best demonstrated with US and the sonographic appearance of a septated cystic mass with clear fluid is supposed to be characteristic of a LA.

However, preoperative diagnosis is often difficult, as in our case because cysts were not clearly defined whilst mainly lacunar spaces were appreciated [2, 4]. The administration of intravenous contrast medium could have better demonstrated the multicystic structure of the mass thanks to the enhancement of the cyst walls and septa, but we were not arranged to perform contrast enhancement US in our institution. Nevertheless, elastosonography was useful to reveal the soft consistency (green color) of the mass and to demonstrate that hypoechoic lacunar spaces, although not clearly cystic, had a fluid consistency because appeared red on colorimetric map of tissue hardness. However, LA elastosonographic findings have not yet been reported, to our knowledge. To ascertain the origin of the cystic mass, characterize its contents and show the extent of cyst involvement, CT and Magnetic Resonance Imaging (MRI) are useful employed. Although CT is burdened by the use of ionizing radiations it is faster and more feasible than MRI when one should avoid sedation. LA typically appears as a large, thin-walled, unilocular or multicystic mass. Intravenous contrast-enhanced CT/MRI may show enhancement of the cyst wall and septa. On CT images, fluid content is typically homogeneous and its attenuation ranges from that of water to that of fat, showing negative attenuation values when chyle is present. Calcifications may occur, but are uncommon [2, 4–6]. In our case, CT images did not show large cystic cavities, however were appreciated low-negative attenuation areas, due to fatty-chylous component, and some calcifications also. The differential diagnosis mainly includes the other mesenteric cystic lesions, such as multicystic mesothelioma, however, because there are no specific radiologic features and its presentation is unspecific, differentiation requires histological evaluation [1, 5, 7]. The treatment of choice is complete surgical excision. The long-term prognosis is favorable if complete excision has been performed. Otherwise the child should be followed up with serial imaging examinations to exclude recurrence of the lesion.

Another treatment reported is image-guided percutaneous catheter drainage of LA followed by sclerotherapy [4]. In our case, patient underwent complete surgical excision and about a month after the operation, the patient was brought for follow-up and did not manifest postoperative complications.

Conflict of interest Raffaella Capasso, Eugenio Rossi, Massimo Zeccolini, and Antonio Rotondo declare that they have no conflict of interest regarding the publication of this article.

Informed consent All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2000. All patients provided written informed consent to enrolment in the study and to the inclusion in this article of information that could potentially lead to their identification.

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