Mesenteric Lymphangioma in the Pediatric Ages

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Received: February 20, 2019; Published: March 21, 2019

Abstract

Introduction: Within the range of congenital malformations that surgeons have to deal with, vascular tumors and malformations represent an special group of entities that are still under study.

Objectives: To determine the clinical-surgical characteristics of a series of two cases with congenital malformations of lymphatic origin.

Cases Presentation: Two patients of 12 and 4 years old respectively that were admitted in the Emergencies services in the Clinica-Surgical Hospital of Guayaquil, Ecuador, with manifestations of intestinal obstruction and abdominal cystic tumoral lesions that required surgical treatments with different techniques. Final evolution was satisfactory.

Conclusion: Intestinal lymphangioma are part of the kinds of clinical presentations of lymphatic origin’s malformations. These have a low incidence but due to their localization and symptoms can require emergency surgical treatment. Their evolution and resolution is satisfactory due to the low index of recidive after complete exeresis.

Keywords: Mesenteric Lymphangioma; Abdominal Cystic Tumor; Pediatrics

Introduction

Within the spectrum of congenital malformations with which a pediatric surgeon must contend, tumors and vascular malformations undoubtedly represent a special group of entities on which medical studies are currently being carried out. Lymphatic malformations are part of these conditions and constitute a form of presentation at the level of the lymphatic vascular system [1].

The lymphangiomas constitute a congenital malformation of benign character with an incidence of 6% within the rest of the benign tumors. Although their location is varied, these are most frequently located in the cervical region 75%, followed by the axillary region in 20% of cases [2]. The abdominal location represents 5% of less frequent presentations and within this the mesenteric is the most common [3]. The location in solid organs has been published as rare in the splenic tissue, liver, bile duct and lung [2,4].

Given their low incidence and the benign nature of these lesions are sometimes classified as unknown, but their real importance lies in the probability of causing abdominal complications such as bleeding, occlusive symptoms, recurrent abdominal pain and intestinal perforations. The first description of a mesenteric cystic tumor is made in 1507 by Beneviene cited by Fontirroche [3].

Three centuries later Tillaux reports the surgical treatment of the first cases in history. From the first reports it is considered that more than 820 cases have been published in the medical literature [5].

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The etiology of these lesions continues to be considered idiopathic, although there are different theories about their development where the congenital origin is considered with greater force.

Based on the above, the objective of this study was to determine the clinical and surgical characteristics of our series of two cases and compare them with the updated international medical literature.

Presentation of Cases

Case 1

A 12-year-old male patient who came to the emergency department with abdominal pain to colic type of about 24 hours evolution, associated with vomiting and abdominal distension. Physical examination revealed a more pronounced distended abdomen in the inferior hemiabdomen, painful on palpation. The radiographic studies showed signs of intestinal occlusion, so their surgical exploration was decided. Laboratory tests within normal limits.

Surgical description: Cystic tumor that occupies the root of the mesentery of the ileal jejunum junction. Complete exeresis of the cystic mass is performed respecting the mesenteric vascular arches at this level. Satisfactory postoperative evolution (Figure 1).

Case 2

Male patient, 4 years of age, who went to the emergency department with abdominal pain of 12h of evolution of colic type associated with vomiting that did not yield with medication. Clinical examination showed signs of moderate dehydration, abdomen distended in a uniform manner; painful on palpation, no visceromegaly, decreased hydro-aerial sounds. Empty rectal ampule. Laboratory studies showed no signs of infection, without other alterations. Radiological studies: Simple abdomen of foot; alterations were observed in the distribution of the gas pattern with a distended loop image in the upper abdomen and air-fluid level in serial studies. Evolutionarily the clinical picture remained so it was decided surgical treatment.

Surgical description: Multilobulated cystic tumor that occupies the root of the mesentery at the jejunal ileal junction in all its thickness. It was decided to perform excision of the tumor with resection and primary terminal-terminal anastomosis of the small intestine. Satisfactory postoperative evolution (Figure 2).

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Discussion and Conclusion

Mesenteric lymphangioma as a lymphatic vascular lesion has similar characteristics to extra-abdominal locations and its actual clinical behavior depends precisely on its location.

The diagnosis of these tumors can be made incidentally from the neonatal stage by imaging studies. It is published that usually 60% of patients are diagnosed during the first 5 years of life; while the remaining percentage can pass to the adult stage [6]. The abdominal location of the lymphangioma is not among the most frequent. However, during the pediatric age this can be present in about 45% of cases. The involvement of the small bowel mesentery is recorded as the most frequent site with 50 - 67% [7] followed by the involvement of omentum, mesocolon and retroperitoneum [8,9].

The literature reports a higher frequency of males with a 3/2.3 relationship. Both aspects: location and presentation by sex are coincident with the results of our patients.

Although it is suggested that the etiology of these lesions is idiopathic, their congenital origin is accepted within the theories of their development. Recall that the lymphatic system develops from the fifth week of the embryonic period with the formation of six lymphatic sacks and their subsequent development depends on the intercommunication of these with the vessels and lymphatic capillaries in different areas of the body.

The appearance of this malformation is then explained by an involvement in the communication of the lymphatic vessels or their agenesis in the fetal stage. The non-interconnected lymphatic vessels dilate until the formation of the final tumoral cystic structure [10]. Other proposed theories are related to alterations in endothelial permeability and lymphatic obstruction [2].

In the medical literature, several nomenclatures have been used for the study of mesenteric cysts where abdominal lymphangiomas have been analyzed. One of the most used is the one developed by Perrot [11] where this lesion is already considered typical of the pediatric age. Similarly, medical studies have already pointed out the importance of considering mesenteric lymphangiomas as distinct from mesenteric cysts originating from mesothelial tissue, given the variations in their clinical behavior [12].

The clinical presentation of abdominal lymphangiomas is closely related to several factors, especially anatomical ones. The location of the tumor in the first instance will be significant because the mesenteric location will most likely affect the involvement of the vascular root of the intestine and its permeability. In as much, the locations in free organs as the omentum, the tumor will need a greater growth to produce compressive effect on the adjacent organs.

In this sense, the dimensions of the tumor will be another factor that will condition the initiation of the clinical manifestations, which will be closely related to the age of the patient given the structural differences of the intra-abdominal organs in the early stages of life.

Figure 1: Patient of 12 years trans-operative.

our patients, this was a characteristic that defined the difference in the surgical technique developed to perform the exeresis of the tumor in each case.

The presence of the tumor can be asymptomatic in up to 50% of cases [7] and can go on to advanced stages of life or be diagnosed incidentally by imaging studies or by increasing the abdominal perimeter. A percentage of the cases present with a recurrent abdominal pain that can be established as an acute abdomen in certain circumstances [13,14]. Symptomatology is usually related to a mechanical obstructive syndrome given the compression, angulation or volvulation of the intestine adjacent to the tumor [2,15] although patients with bleeding pictures due to erosion of the walls of the cystic tumor have been reported.

The compression of the urinary tract with renal infectious involvement can be related to retroperitoneal tumor locations [3], while the symptomatology of the locations in solid intra-abdominal organs will be manifested by the presence of the tumor per se or functional alterations of the organ in question.

The development of imaging techniques to date has allowed the diagnosis of these cystic tumor lesions from the fetal stage. The use and complementation of an abdominal ultrasound, computerized axial tomography (CT) and magnetic resonance provide us with enough data to reach a preoperative diagnostic conclusion.

The ultrasound vision of the lesion will correspond to a hypoechoic lesion with septa or septa inside it; the echogenicity will vary depending on the intracystic content. The CT will give us better elements of extension and involvement of neighboring organs, as well as allow us to assess the intracystic content indicating the degree of benignity.

The use of simple abdominal radiographs will be more associated with the diagnosis of the abdominal pain in its acute phase, being able to find signs of intestinal occlusion and radiopacity of an abdominal area with changes in the distribution of the gas pattern. Also intra-abdominal calcifications could be defined with this medium.

The use of contrasted radiological studies of the digestive or urinary tract may be useful in some particular case [16].

The treatment of abdominal lymphangiomas is surgical and its objective is to perform the exeresis of the tumor. The technical variants to be used will depend on the anatomical characteristics of the lesion and the involvement of neighboring organs. So that, whenever possible, the simple exeresis of the lesion will be performed and in other occasions it will be necessary to perform intestinal resections with primary anastomoses associated with the tumor extirpation. A final option would be marsupialization of the lesion to the cavity when the exercise option is not possible. This would never be a good solution given the high rate of recurrence of lymphangiomas.

The diagnosis of intratumoral infection will be to some extent a condition for deferring surgical treatment, as long as there is no involvement of the abdominal cavity [17].

The treatment options applied to lymphangiomas located in soft tissues are not useful in the intra-abdominal location or have been replaced due to the poor results of the use of laser; drainage and aspiration [18]. On the other hand, the injection of intracystic substances such as OK 432 (Picibanil) have up to 10 - 15% of recurrence [19,20].

The approach with minimally invasive surgery could be considered in the surgical treatment of these lesions taking into account their own characteristics, or as a more diagnostic method. As background there are references on this type of treatment variant in mesenteric sites in 1991 by Mackenzie [7] and splenic since 2001 by Know [21]. Currently, different authors have standardized this behavior [2,11].

The final diagnosis as in all lesions by general order will be made from the anatomopathological point of view [22].

From the histological point of view, lymphangiomas are classified into three types: simple, cavernous and cystic. In some publications, variants of lymphangiohemangioma and lymphangiosarcoma may be added to this classification [23].

By nature, from the macroscopic point of view, these malformations are cystic with their macro- or microcystic variants. Its content is usually similar to milky or clear straw-colored lymph, but it could become hemorrhagic [24].

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At microscopic examination, simple or capillary lymphangioma is located on the skin and is composed of thin-walled lymphatic vessels. The cavernous lymphangioma is composed of dilated lymphatic vessels, with a lymphatic stroma and is connected to normal adjacent lymphatic vessels. Finally, the cystic variety consists of lymphatic spaces of various sizes, flat endothelial epithelium whose wall contains thin fascicles of smooth muscle and has no connection with the adjacent normal lymphatic vessels [23,25].

After the review of our patients and the medical literature we could conclude that intestinal lymphangiomas are part of the range of clinical presentations of malformations of lymphatic origin, with a low incidence but, given their location and symptoms, may require surgical treatment of urgency. Definitely its evolution and resolution is satisfactory, due to the low rate of recurrence after the complete exeresis.

Conflicts of Interest
The authors declare that there are no conflicts of interest of any kind.

Bibliography

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Volume 8 Issue 4 April 2019
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