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Editorial Note

Abdominal Lymphangioma in Children: A Pediatric Retrospective Chart Review of 16 Cases

Villalon G, Weissenstein A, Luchter E, Bittmann S

Department of Pediatrics, Ped Mind Institute (PMI), Gronau, Germany

*Corresponding Author: Stefan Bittmann, Head of the Department of Pediatrics and Ped Mind Institute (PMI) Pediatrician, Hindenburgring, Gronau, Germany.

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Abstract

Lymphangiomas are benign lesions of vascular origin that show lymphatic differentiation. They occur in many anatomic locations. Complete resection is difficult in some cases and recurrences are common. A pediatric chart review was carried out in 16 cases. The authors summarized their experience of abdominal lymphangiomas in children.

Keywords: Lymphangioma-abdomen-children

Introduction

Abdominal cystic lymphangioma is a very rare congenital tumor of lymphatic origin. It usually appears in the pediatric age and frequently presents with non-specific symptoms and deceptive signs causing, at times, diagnostic dilemmas.

The diagnostic modalities of choice are ultrasonography and computer tomography imaging. The majority of abdominal lymphangioma are discovered in the early years of life. There are found two major revelation lodes, a tumoral syndrome with pain and a complication like torsion, infection, bleeding into or rupture of the cysts. The management of lymphangioma in children is challenging because complete resection is difficult to achieve in some cases. Recurrences are common. Risk factors for recurrence included location, size, and complexity of the lesions. Spontaneous regression is infrequent and is more often seen with recurrent lesions. In this study we reviewed our experience with abdominal lymphangioma in 16 children.

Classification and Histology

The classification of lymphangiomas is inconsistent in the literature and includes many different criteria. In 1877, Wegner first classified lymphangiomas as simple, cavernous, and cystic [6]. Later, Watson and Mc Carthy added cellular and diffuse systemic lymphangiomas to this classification [7]. Simple lymphangioma is often called lymphangioma simplex or

capillary lymphangioma. It is composed of superficial, scattered channels that give rise to small, shiny, vesicular lesions [7]. Cystic lymphangiomas are large, thin-walled endothelium lined cysts derived from lymphatic tissue [7]. Cellular lymphangioma is characterized by an abundance of proliferating endothelium and clinically gives rise to circumscribed fibrous lesions [7]. The diffuse systemic lymphangioma is a large slow-growing tumor that may be associated with Klippel-Trenaunay-Weber syndrome [8]. This tumor frequently involves an entire extremity, causing hypertrophy with overgrowth of the entire extremity [7]. In cases, where a clear histological diagnosis cannot be achieved, immunohistochemical examinations have to be done.

Results

The diagnosis of abdominal lymphangioma was confirmed in 16 patients. A retrospective pediatric chart review carried out 9 boys (56.3%) and 7 girls (43.7%). Histocytological confirmation of lymphangioma was obtained in all patients. Abdominal pain was found as initial symptom in 12 cases (75%). In two patients, ultrasonography because of routine check-up for chronic hepatitis B and routine control for recurrent urinary infections revealed an intraabdominal tumor (12.5%). In one patient the tumor was found by intrauterine ultrasonography (6.25%). In one patient only a swelling of the lower abdomen without pain was present (6.25%). Involved sites were the ileum, 5 patients (31.3%); jejunum, 3 patients (18.8%); omentum majus, 2 patients (12.5%); jejunum/ileum, 1 patient (6.25%); mesenterium, 2 patients (12.5%); retroperitoneal and bursa omentalis, 2 patients (12.5%); and combined abdominal, mediastinal, retroperitoneal and the right inguinal channel in 1 patient (6.25%). Of all cases with abdominal lymphangioma, 13 (81.3%) were intraperitoneal and 2 (12.5%) retroperitoneal. In 1 patient an intra- and retroperitoneal location was found (6.25%). Complications included pancreatitis in 3 cases (18.8%) and ileus due to ileoileal invagination in 1 case (6.25%). Age at treatment ranged from 11 weeks to 9 years (median 4.9 years). Treatment consisted of macroscopically complete excision in 12 patients (75%). A partial non-contact laser application therapy (Nd:YAG laser) was performed in 4 patients (25%). A planned complete excision of the lymphangioma was performed in 1 patient (6.25%). This patient was diagnosed after a routine thorax x-ray with subsequent work-up including US and MRI of the thorax and abdomen. In 3 patients a segmental small bowel resection was performed by the surgical unit. (18.8%). In one patient (6.25%), because of organ involvement, a pancreas tail resection was performed. No recurrences were found after treatment. Spontaneous regression was never found.

Discussion

In 1828, Radenbacker described the first case of a cystic lymphangioma [9]. Abdominal lymphangioma in childhood is a rare tumor of the mesenteric and/or retroperitoneal lymphatics [16]. Abdominal lymphangioma has an incidence of less than one per 100.000 hospital admissions [4]. They are rare, accounting for less than 1% of all lymphangiomas. It is found a preponderance in males with a male:female ratio of 3:1 [4,10]. As many as 90% may manifest before the age of three [12].

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Table 1: Pre-and postoperative variables of 16 children with abdominal lymphangioma.

Patient	Symptoms	Localization	Weight / size	Histology	Age at surgery	Postop course
1	Abdominal pain	Jejunum	450gr; 12 x 10 x 6 cm	Cystic lymphangioma	7y	Pancreatitis, high lipase
2	Abdominal pain	Retroperitoneum, and bursa omentalis	95.2gr; 9.5 x 7.5 x 5.5cm And: 3.5 x 2.5 x 1.5cm pancreas tail (11.7gr)	Cystic lymphangioma	9y	High amylase
3	Abdominal pain, vomiting	Ileum	No information		4.5y	No compl
4	Recurrent abdominal pain since 6 months	Omentum majus	No information	Multicystic lymphangioma	9y	No compl
5	Vomiting since 2 weeks, abdominal pain	Ileum and jejunum	1.6 x 2.7cm	Multicystic mesenterial lymphangioma, lymphangiektasia of the small bowel	11 weeks	No compl
6	Abdominal pain since 2 weeks	Mesenterium of colon ascendens, descendens, sigma and rectosigmoideal region, whole retroperitoneum,, mediastinal, right inguinal channel	Diffuse manifestation	Multicystic lymphangioma, reactive sinushistiocytosis	3y	No compl
7	Abdominal pain,	Omentum majus	221gr; 11x8 x 6.5cm	Multicystic lymphangioma	2y	No compl
8	Recurent urinary infections: US reveals intraabdominal TU	Ileum	222gr,; 11.5 x 9 x 5cm	Multicystic lymphangioma	7y	Postop ileus due to ileo-ileal invagination of 10cm, manual desvagination
9	Abdominal pain, obstipation	Ileum	No information	Cystic lymphangioma	3y	No compl
10	Vomiting,, chronic hepatitis B: US reveals intraabdominal TU	Ileum	8 x 6 x 3cm	Cystic lymphangioma with small intramural cavernous lymphangioma	4y	No compl
11	Abdominal pain	Retroperitoneum and bursa omentalis	10.5 x 6.5 x 4.5 cm	Cystic lymphangioma	7 y	Pancreatitis, high lipase
12	Intrauterine diagnosed TU	Mesenterium of he whole small bowel	No information	Cystic lymphangioma	4 mo (partial excision, non contact laser application)	No compl
13	Swelling of the lower abdomen found by school doctor	Mesenterium of jejunum	No information	Cystic lymphangioma	4 y	No compl
14	Abdominal pain, vomiting	Mesenterium of ileum	No information	Cystic lymphangioma	13y, partial excision, marsupialisation	No compl
15	Abdominal pain	Mesenterium of jejunum	58.3 gr,	Multicystic lymphangioma	4y , partial excision of jejunum	No compl
16	Abdominal pain	Whole mesenterium		Cystic lymphangioma	4 mo, partial excision, non contact laser application	No compl

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They are considered to be congenital malformations stemming from sequestration of lymphatic tissue. On the other hand Godart postulated that premature lymphatics appear as mesenchymal slits, which coalesce and normally communicate with the venous system [11]. Failure of establishing this communication may lead to lymphangioma. Both theories would explain, why lymphangiomas affect young children and are preferentially located at sites where lymphatic sacs occur. The presentation of abdominal lymphangioma is more acutely in children compared to adults [1]. In adults the history seemed to be longer and tumors were more often found in the retroperitoneum [1]. The lesion differs from a mesenteric cyst as it follows a proliferative and invasive course and the different histology of the endothelium [4]. Lymphangiomas discovered fortuitously or during a complication in the early years of life. Two major revelation lodes are found: a tumoral syndrome with pain or a complication like torsion, infection, bleeding or rupture. Clinical presentation can be misleading. The clinical picture is often pleomorphic and noncharacteristic: therefore, complex imaging studies are necessary in the evaluation of this condition. US and CT have a major role in the correct preoperative diagnosis and provide important information regarding location, size, adjacent organ involvement, and expected complications [14,15]. The intra- or retroperitoneal location of the lesion is sometimes difficult to determine by sonography, in which case CT scanning is usually adequate. Multisliced computed tomography with its excellent three dimensional imaging capabilities and ability to visualize vessels is ideally suited for evaluation of patients with suspected abdominal lymphangioma, where a rapid and accurate preoperative localization of the lesion is critical for facilitating proper management [13]. Only few cases are reported in which a laparoscopic excision [17-22] or a laparoscopic interstitial laser treatment with Nd:YAG laser [19] of the lymphangioma was successfully performed. Extensive studies concerning this treatment modalities were not found in the literature. In conclusion we reported the pediatric chart experience of 16 cases of abdominal lymphangioma. The treatment of choice for abdominal lymphangiomas is complete surgical treatment. As they are diffuse, total excision may require resection of involved abdominal organs and bowel to prevent recurrence. Recurrence can be found in cases with incomplete resection of the lymphangioma. Early recognition and appropriate treatment of these lesions are associated with a good prognosis [5].

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