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Abdominal lymphangiomas in adults: case report and literature review

Suaugusiųjų pilvinės limfangiomos: klinikinis atvejis ir literatūros apžvalga

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Intraabdominal lymphangioma is rare and comprises 5% of all lymphangiomas. In adults, it is less common than in children, and because of a benign progress lymphangioma is usually asymptomatic and its diagnosis is quite difficult. However, it is recommended to extirpate lymphangioma surgically to avoid complications. There is the question if lymphangioma should be completely excised to avoid relapses. We present a case of cavernous lymphangioma in an adult male who underwent a partial resection of the tumour. After ten years of observation, there is no evidence of tumour recurrence.

Key words: lymphangioma, cavernous, cystic

Pilvinės limfangiomos pasireiškia retai ir sudaro mažiau nei 5 % visų limfangiomų. Suaugusiesiems limfangiomos pasitaiko rečiau nei vaikams, dėl gerybinės eigos jos būna besimptomės ir sunkiai diagnozuojamos. Vis dėlto siekiant išvengti komplikacijų rekomenduojamas chirurginis limfangiomų šalinimas. Literatūroje išlieka diskusinis klausimas, ar būtina visiškai pašalinti limfangiomas siekiant išvengti jų atkryčio. Mes aprašome kaverninės limfangiomos atvejį vyrui, kuriam buvo atlikta dalinė rezekcija. Po dešimties stebėjimo metų atkryčio nerasta.

Reikšminiai žodžiai: limfangioma, kaverninė, cistinė

Introduction

Lymphangioma is defined as a congenital malformation of the lymphatic system [1]. Lymphangiomas are classified into three histologic types: capillary, cystic, and cavernous. The capillary type is usually found in the skin (mostly in the neck, head and axillary region) [2, 3]. Cystic and cavernous lymphangiomas usually are intraabdominal – 5% of all diagnosed [4]. They are usually found in the retroperitoneum (23% of intraabdominal lymphangiomas according to some authors) and mesentery (70% of intraperitoneal lymphangiomas) [5, 6]. Lymphangiomas in the pancreas are extremely rare.

There are only about 70 reported cases of pancreatic lymphangiomas in the literature [7].

Typical diagnostic methods for lymphangiomas are ultrasonography (US) and computed tomography (CT). The features of cavernous lymphangioma in US and CT imaging are a multilocular homogeneous mass, enhancement of the wall and septa, and the presence of a fluid [8–10] (Figures 1, 2). Calcification in lymphangiomas occurs rarely [5] (Figure 3). CT and US are useful in detecting the cystic nature, size and location of the lesion [9]. On CT and US imaging, a cavernous lymphangioma may appear as a solid mass due to the microcystic nature, intracystic debris, or haemorrhage. In these cases, magnetic resonance imaging (MRI) is useful as it is more specific for detecting the cystic nature of cavernous lymphangioma [10]. MRI T1 weighted images show heterogeneous lesions, while T2-weighted images indicate cystic spaces filled with a fluid [8].

However, the diagnosis of intraabdominal lymphangioma is quite difficult as it is usually asymptomatic [2, 11]. The most common symptom of lymphangioma is abdominal pain [12]. In some cases, lymphangioma can cause gastrointestinal bleeding [2]. The main treatment choice for lymphangiomas in the abdomen is surgery. Radical extirpation is recommended as incomplete excision gives a risk of recurrence. However, there are no described cases of malignant degeneration in the literature. Surgical treatment should be as urgent as possible, thus the complications of lymphangiomas can be avoided. These complications include infection, haemorrhage, bowel obstruction, ascites, and the growth of the tumour [6]. Therefore, we would like to present a case of cavernous lymphangioma and our experience in treating lymphangiomas.

Case report

In ten years of our experience, there were six cases of lymphangiomas in our center (Table). There were 4 male and 2 female patients with an average age of 29 ± 9 years (range, 20–44). The main clinical symptom was abdominal pain (n = 4). Somewent patients had other symptoms: icterus, frequent urination, and back pain. In one case, lymphangioma was asymptomatic. All patients were examined with the abdominal US and

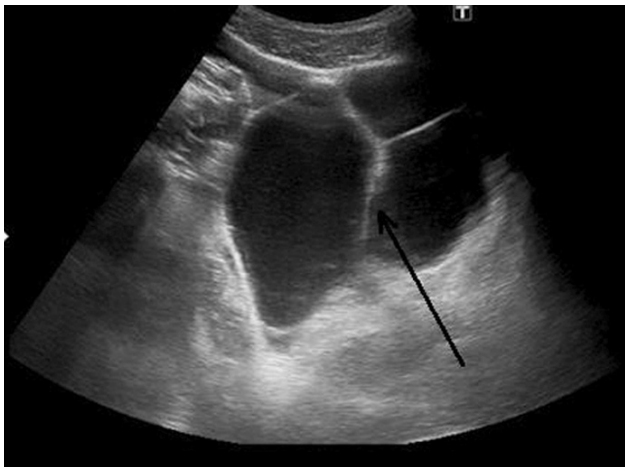

Figure 1. Arrow showing septa of the cystic mass in US

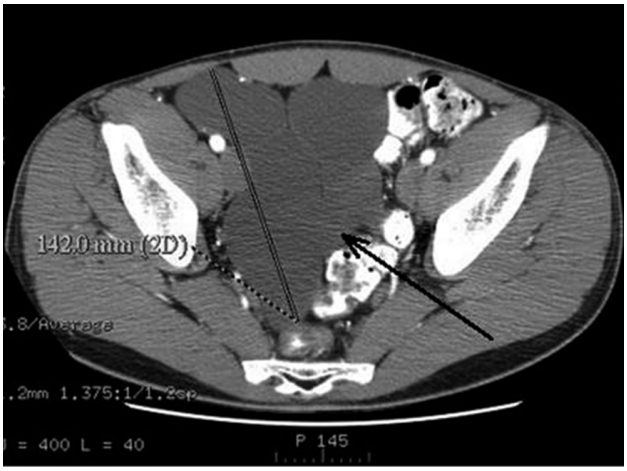

Figure 2. Arrow showing multilocular homogeneous mass in the abdomen and retroperitoneum revealed in a CT scan

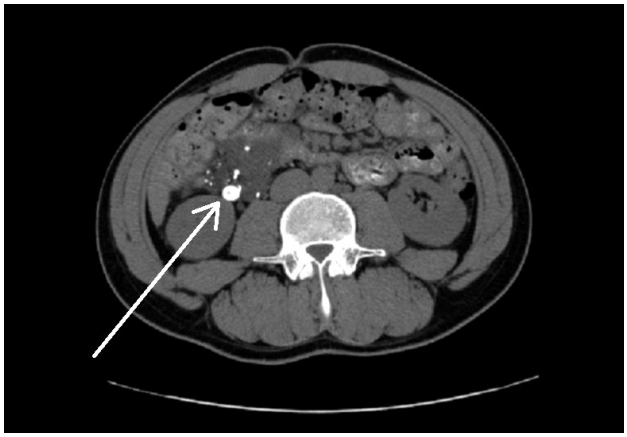

Figure 3. Arrow showing tumour calcification found in the CT scan

Table. Characteristics of patients with lymphangiomas

Patient	Age	Sex	Symptoms	Size of the tumour (cm)	Surgery	Localization and involvement of surrounding tissues	Histology	Length of follow-up (years)
1	20	Male	asymptomatic	23 × 13 × 7	Laparotomy, PDR, extirpation of the tumour	Pancreas, greater omentum	Cavernous lymphangioma	3
2	34	Male	Right abdominal pain	9 × 7.8 × 7	Laparotomy, PDR, extirpation of the tumour	Pancreas, peripancreatic adipose tissue, stomach and duodenum	Cavernous lymphangioma	3
3	20	Male	Right abdominal pain, icterus	7 × 3.7 × 5.7	Laparotomy, resection of the tumour, stenting of the common bile duct	Mesentery	Cavernous lymphangioma	9
4	26	Female	Abdominal pain, frequent urination	15 × 9 × 4	Laparotomy, extirpation of the tumour	Mesentery	Cavernous lymphangioma	2
5	29	Male	Back pain	9.5 × 5 × 11	Laparotomy, PDR, extirpation of the tumour	Pancreas	Cavernous lymphangioma	6
6	44	Female	Right abdominal pain	6 × 3 × 2	Laparotomy, extirpation of the tumour	Mesentery	Cystic lymphangioma	1

PDR – pancreaticoduodenal resection.

CT scan. The preoperative diagnosis was made through typical radiological images. All patients underwent surgical procedures with the further histopathological analysis. In 50% of cases, lymphangiomas were mesenteric (n = 3), while in other 50% cases the localization of lymphangiomas was in the pancreas (n = 3). In three cases, pancreatoduodenal resection with radical tumour extirpation was made, in two cases radical extirpation of the mesenterium tumour and in one case the resection of the tumour were performed. In the latter case, part of the tumour was left in the hepatic hilum because of the technical difficulties to extirpate it. No relapse of lymphangioma was observed in any case. Histopathological diagnosis in 5 cases was cavernous lymphangioma, and there was one case of cystic lymphangioma. Patients were followed up once per year with CT and the other year with US. A patient who has been followed up for 9 years undergoes CT every 3 years.

In 2003, a 20-year-old Lithuanian man who complained of abdominal pain and icterus underwent surgery of the retroperitoneal tumour above the right kidney.

Partial resection of the tumour and cholecystectomy were performed. The histologic results showed lymphangiomatosis. During the observation of the patient, the level of total bilirubin was always above 40 μmol/l, although the patient did not complain of the pain and the icterus did not progress. After three years, the symptoms (right abdominal pain and icterus) recurred. As these symptoms were continuing, he went to the hospital. Abdominal US and CT showed tumour in the hilum of the liver. In 2005, the patient was operated on due to the tumour in the mesentery. During the surgery, the duodenum was mobilised (the Kocher manoeuvre). A common hepatic artery, a hepatic portal vein, and a common bile duct were separated, the tumour in the

ligament was extirpated, and the lymph nodes (group 13 and group 8) were excised. In the hilum of the liver, part of the tumour was left due to the technical difficulties to extirpate it.

During the surgery, a bile duct was opened through the stump of the cystic duct, which was prepared with a stent. At the end of the operation, bile ducts were stented with internal stents, and the area of choledochotomy was stitched. Four days after the operation, the bile began to flow through the drain. During the endoscopic retrograde cholangiopancreatography (ERCP), the bile flowing through the stent was seen. After elimination of the stent, a leakage was observed; however, restenting was not successful. Therefore, repalarotomy was performed. During the operation, the bile ducts were restented and the common bile duct was stitched. The further postoperative course was fluent. The histologic results showed a fibrous adipose tissue with nerve trunks and a large amount of small and dilated vascular cavities filled with erythrocytes. There were also four lymph nodes with reactive alterations – follicular hyperplasia and sinus histiocytosis.

The patient underwent abdominal US every six months to check relapses. The last abdominal US showed a 5.7 × 2.5 cm fibrous tissue in the area of the hepatic hilum (Figure 4).

After ten years of observation, there were no recurrence, no complaints and icterus were observed.

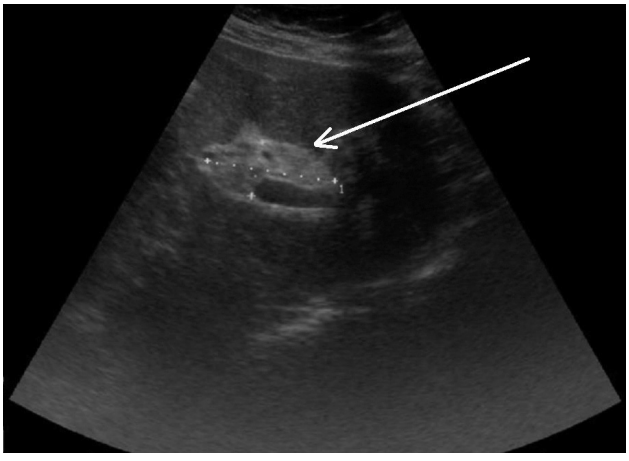

Figure 4. Arrow showing fibrous tissue in the hilum of the liver found by US

Discussion

According to the literature, cavernous lymphangiomas tend to be found in the retroperitoneum or retroperitoneal organs in adults [13]. In our experience, there were two adults with tumour in the retroperitoneal organs, one in the retroperitoneal organs and stomach, and three tumours in the mesentery [4]. However, lymphangiomas in the pancreas are found in less than 1% of all lymphangiomas, in contrast to our experience (50%). Pancreatic lymphangiomas are usually found in women (64% of all cases, according to some authors), while in our cases the male-to-female ratio was 2 : 1 [7].

Lymphangiomas occur mainly in children, and 80–90% of them are diagnosed with a lymphangioma within the first few years of life. In adults, they are often asymptomatic or with nonspecific symptoms, therefore observed late [2]. According to some authors, like in our experience, lymphangioma tends to be diagnosed in young males [13].

The preoperative diagnosis of lymphangiomas is complicated. Diagnostic methods, such as abdominal US and CT, are useful in diagnosing tumour, although cavernous lymphangiomas, which appear on CT as a solid mass, may be confused with metastases in patients with malignancy [2, 10]. In our experience, all patients were diagnosed with a lymphangioma preoperatively. Percutaneous biopsy of the tumour is not advisable because it is of cystic nature and shows a risk of bleeding [9]. Also, in some cases, it is hard to take a biopsy without damaging other organs.

A cavernous lymphangioma consists of dilated lymphatic vessels and a lymphoid stroma. Unlike cystic lymphangiomas, it is connected with the normal lymphatics [10]. The main histological features of a cavernous lymphangioma are single-layer endothelial-lined lymphatic spaces containing chylous or serous material. In our cases, another characteristics of tumour were smooth muscles, eosinophils, and lymphoid folicles [14]. A cystic lymphangioma may be a long-standing cavernous lymphangioma in which cavernous spaces were enlarged [15].

Patients with cavernous lymphangiomas often have anemia [14]. In cases of acute bleeding, angiography and embolization can be applied [16]. However, in our experience, only one patient had anemia. Since it was

caused by chronic bleeding from the lymphangiomas-ulcerated stomach, angiography and embolization were not required.

A complete surgical excision is recommended, as the rate of recurrence after incomplete resection is 40%. On the other hand, even after a radical surgical operation, there may also be a recurrence. Because of that, a follow-up after operation is necessary. According to the literature, lymphangiomas grow very slowly [5]. Because of the slow growth of the tumour, stenting of the bile ducts and sclerotherapy may be also effective treatment options.

In our experience, there were no relapses after both complete and incomplete resection. There is a question if surgery is needed when the growth of a tumour is slow. In our opinion, if the lymphangioma is asymptomatic and with a typical radiological image, observation is sufficient. Thus, postoperative complications can be avoided. After the surgical treatment we recommend the further observation: abdominal CT imaging every 3 years and US every year. As an alternative for CT, MRI may be used for observation.

Abdominal lymphangiomas should be excised completely as they tend to penetrate important structures.

Conclusions

Despite the fact that the preoperative diagnosis of lymphangiomas is difficult, US and CT are reliable diagnostic tools for it. It is recommended to extirpate a tumour and to follow up the patients because of the risk of recurrence. Complete excision of the tumour gives good results, although when it is not feasible, incomplete resection may also be optional. In our opinion, observation is sufficient for patients with a typical radiological view and slow tumour growth.

REFERENCES

1. Giguère CM, Bauman NM, Smith RJH. New treatment options for lymphangioma in infants and children. Ann Otol Rhinol Laryngol. 2002 Dec; 111(12 Pt 1): 1066–1075.
2. Rai P, Rao RN, Chakraborthy SBD. Caecal lymphangioma: a rare cause of gastrointestinal blood loss. BMJ Case Rep. 2013.
3. Matsuba Y, Mizuiri H, Murata T, Niimi K. Adult intussusception due to lymphangioma of the colon. J Gastroenterol. 2003; 38(2): 181–185.
4. Suthiwartnarueput W, Kiatipunsodsai S, Kwankua A, Chaumrattanakul U. Lymphangioma of the small bowel mesentery: a case report and review of the literature. World J Gastroenterol WJG 2012 Nov 21; 18(43): 6328–6332.
5. Makni A, Chebbi F, Fetirich F, Ksantini R, Bedioui H, Jouini M, et al. Surgical management of intra-abdominal cystic lymphangioma. Report of 20 cases. World J Surg. 2012 May; 36(5):1037–1043.
6. Wani I. Mesenteric lymphangioma in adult: a case series with a review of the literature. Dig Dis Sci. 2009 Dec; 54(12): 2758–2762.
7. Dalla Bona E, Beltrame V, Blandamura S, Liessi F, Sperti C. Huge cystic lymphangioma of the pancreas mimicking pancreatic cystic neoplasm. Case Rep Med.; 2012. Available from: http://www.ncbi.nlm.nih.gov/pmc/articles/PMC3502873/
8. Philippakis GE, Manoloudakis N, Marinakis A. A rare case of a giant cavernous lymphangioma of the chest wall and axilla in an adult patient. Int J Surg Case Rep. 2013; 4(2): 164–166.
9. Vennarecci G, Ceribelli C, Laurenzi A, Moroni E, Ettorre GM. Giant cavernous mesenteric lymphangioma in adult. Updat Surg. 2013 Dec; 65(4): 317–319.
10. Hwang SS, Choi HJ, Park SY. Cavernous mesenteric lymphangiomatosis mimicking metastasis in a patient with rectal cancer: a case report. World J Gastroenterol WJG. 2009 Aug 21; 15(31): 3947–3949.
11. De Perrot M, Rostan O, Morel P, Le Coultre C. Abdominal lymphangioma in adults and children. Br J Surg. 1998 Mar; 85(3): 395–397.
12. Chung JC, Song OP. Cystic lymphangioma of the jejunal mesentery presenting with acute abdomen in an adult. Can J Surg. 2009 Dec; 52(6): E286–288.
13. Goh BKP, Tan Y-M, Ong H-S, Chui C-H, Ooi LLPJ, Chow PKH, et al. Intra-abdominal and retroperitoneal lymphangiomas in pediatric and adult patients. World J Surg. 2005 Jul; 29(7): 837–840.
14. Morris-Stiff G, Falk GA, El-Hayek K, Vargo J, Bronner M, Vogt DP. Jejunal cavernous lymphangioma. BMJ Case Rep 2011.
15. Ogun GO, Oyetunde O, Akang EE. Cavernous lymphangioma of the breast. World J Surg Oncol 2007; 5: 69.
16. Chen G, Cui W, Ji X-Q, Du J-F. Diffuse hemolymphangioma of the rectum: a report of a rare case. World J Gastroenterol WJG. 2013 Mar 7; 19(9): 1494–1497.