CASE REPORT

Oesophageal lymphangioma: an exceedingly rare tumour

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SUMMARY

An asymptomatic 57-year-old man was referred for evaluation of a 10 mm pale pink subepithelial polypoid lesion of the distal oesophagus; the lesion was found incidentally during an upper gastrointestinal endoscopy. The endoscopic ultrasound (EUS) revealed a heterogeneous, although primarily hypoechoic, well-circumscribed lesion, located in the submucosa. Owing to the absence of endosonographic features, which would have allowed for a precise diagnosis, EUS-guided fine-needle aspiration was performed; however, the cytology was inconclusive because of the lack of cellular representation. We chose to remove it by cap-assisted endoscopic mucosal resection. However, due to its shape and deep location, complete removal was not achieved. A macrobiopsy was taken, and histological examination, of all oesophageal layers except the adventitia, revealed many irregularly dilated lymphatic vessels, beneath normal squamous epithelium. These findings are consistent with the diagnosis of lymphangioma. A follow-up EUS did not reveal any changes. The patient is currently asymptomatic and is under surveillance.

BACKGROUND

Lymphangiomas are benign soft tissue tumours that are most likely congenital malformations of sequestered lymphatic tissue that fails to communicate with the normal lymphatic system.¹

They have been reported in every organ except the brain. Their most frequent locations are in the head and neck region. Diagnosis is often made in the first year of life (50–60%) and in children younger than 2 years of age (90%).²

Only 1% of lymphangiomas develop in the gastrointestinal tract, with the colon and rectum being the most common sites, followed by the stomach, duodenum, small intestine and oesophagus.³

Only 22 cases of lymphangioma of the oesophagus have been reported worldwide. Owing to the extreme rarity of oesophageal lymphangiomas, the authors present a case of one such tumour, found incidentally during an upper gastrointestinal endoscopy.

CASE PRESENTATION

An asymptomatic 57-year-old man was referred to our department for evaluation of a 10 mm pale pink subepithelial polypoid lesion of the distal oesophagus; the lesion was found during an upper endoscopy that was performed as part of surveillance for peptic ulcer disease (figure 1). The overlying mucosa was apparently normal.

INVESTIGATIONS

The endoscopic ultrasound (EUS) revealed a 12.5 mm×6.5 mm, heterogeneous, although mainly hypoechoic, well-circumscribed lesion, located in the submucosa layer, 35 cm from the incisor teeth; the muscularis propria layer was intact and no mediastinal or coeliac trunk adenopathies were observed (figure 2). Owing to the absence of endosonographic features, which would have allowed for a precise diagnosis, EUS-guided fine-needle aspiration (EUS-FNA) was proposed. EUS-FNA was performed a few months after the first examination and showed a slight increase in the size of the subepithelial lesion. Cytological findings were inconclusive due to lack of cellular representation.

TREATMENT

Taking these findings into account, we chose to remove the lesion by cap-assisted endoscopic mucosal resection in the operating room. However, due to the shape and deep location of the lesion, complete removal was not achieved. A macrobiopsy was taken, without complications. The histological examination of all oesophageal layers except the adventitia, revealed several empty, thin-walled, tortuous, irregularly dilated lymphatic vessels of varying sizes lined by a single layer of flattened endothelium, beneath normal squamous epithelium (figure 3). These findings are consistent with a diagnosis of lymphangioma. There was no evidence of dysplasia or malignancy.

OUTCOME AND FOLLOW-UP

A follow-up EUS did not reveal any changes in the lesion. The patient is currently asymptomatic and is under surveillance at our department.

DISCUSSION

Lymphangioma of the oesophagus is an extremely rare entity.⁴ It was first described by Watson-Williams in 1934.⁵ A MEDLINE literature search identified only 22 cases of oesophageal lymphangioma (table 1), and we report the 23rd case.

The average age at diagnosis of oesophageal lymphangioma is 57 years (range between 32 and 72 years), and it predominantly occurs in men. These tumours are usually small, with almost half of those reported under 2 cm in diameter, as in the current case; however, two large lesions (10×2.5×1 and 6×5×4 cm) were recently described by Xue *et al.*²⁵ Oesophageal lymphangiomas are almost always located in the distal portion of the oesophagus. Although solitary lesions are the

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Figure 1 Endoscopic view of the subepithelial polypoid lesion in the distal oesophagus.

most common by far, there have been three reports of oesophageal lymphangiomatosis.^{17 20 25}

The clinical presentation of this entity varies, depending on the location and size of the tumour.²³ The vast majority of patients are asymptomatic, and the tumour is incidentally discovered during endoscopic investigation for symptoms not related to it,⁸ as in the current case.

On endoscopic examination, an oesophageal lymphangioma appears as a pale, pink, white or yellowish translucent subepithelial polypoid mass projecting into the lumen, and covered with normal mucosa; it is compressible when probed with biopsy forceps.^{4 18 23}

Endoscopic biopsy is usually unrevealing, as these tumours are submucosal and covered by normal squamous epithelium.⁴ Moreover, biopsy should not be performed when the lesion resembles an oesophageal varix.¹⁰ In our case, considering that the lesion was undoubtedly subepithelial, a biopsy was not taken and we initially chose to characterise the lesion by EUS.

Differential diagnosis from other gastrointestinal mesenchymal tumours is mandatory, as some gastrointestinal soft tissue tumours can undergo malignant degeneration.^{13 19}

EUS is the most useful examination for distinguishing lymphangioma from leiomyoma and other subepithelial tumours because it accurately shows the cystic nature of lymphangioma

Figure 2 Endoscopic endosonographic image showing a submucosal tumour with heterogeneous internal echo and intact muscularis propria layer.

Figure 3 Photomicrographs of the resected specimen revealing many irregular dilated lymphatic vessels of varying sizes and overlying normal squamous epithelium (H&E stain).

and whether the oesophageal muscles are intact, which is necessary for the tumour to be amenable to endoscopic resection.^{4 15 18 23} Although a multicystic pattern within the third layer or submucosa is considered a feature of lymphangioma,⁴ not all lymphangiomas have this internal echo structure,^{4 23} because the tumour structure may vary in accordance with the size of the dilated lymphatic vessels.⁴ This is most likely the reason for the absence of the typical endosonographic pattern in our case.

Therefore, definitive diagnosis of oesophageal lymphangioma requires histological examination. Oesophageal lymphangiomas are characterised by thin-walled, dilated lymphatic channels lined by flattened endothelium, which can be empty or filled with chylous or serous fluid,²⁵ and which may contain lymphocytes and, occasionally, erythrocytes.¹⁹ Additionally, these tumours are frequently surrounded by lymphocytic aggregates.²⁶ In our case, histopathological evaluation revealed lymphatic vessels with typical features of lymphangioma. No lymphocytic aggregates were present in the stroma. In most cases, establishing the correct diagnosis based on histological examination is straightforward; however, lymphangiomas with secondary haemorrhage can be confused with cavernous haemangiomas.¹ In the current case, all dilated lymphatic spaces were empty (no erythrocytes were seen in the lumen) and several smaller, rounded, regular and non-ectatic spaces filled with erythrocytes corresponding to normal blood vessels were present in the submucosa, and these features support the diagnosis of lymphangioma over haemangioma.

Because malignant transformation of oesophageal lymphangioma has never been reported in the literature, the management of this tumour can be conservative if the patient is asymptomatic and if a histological diagnosis has already been established.^{18 23}

The presence of symptoms related to the tumour and a significant increase in size or change in configuration of the tumour without a definitive diagnosis of lymphangioma are indications for resection.^{19 23} None of these features was present in our case, so a conservative strategy was adopted.

Endoscopic resection is an appropriate, safe and minimally invasive option for removal of oesophageal lymphangiomas under 2.5 cm in size.^{4 15 18} Large submucosal oesophageal lymphangiomas with a flat configuration are best managed by endoscopic submucosal dissection, as recently reported by Arashiro *et al.*²³ Surgical treatment is necessary when the

Table 1 Cases of oesophageal lymphangiomas reported in the literature

Case	Author	Year	Age	Gender	Symptoms	Site	Size (cm)	Treatment	Reference
1	Watson-Williams	1934	61	Male	Chest pain, vomiting, dysphagia	L	Unknown	Follow-up	5
2	Schmidt	1961	Unknown	Unknown	None	Unknown	Unknown	Postmortem examination	6
3	Brady	1973	62	Female	Epigastric pain	L	5	Follow-up	7
4	Armengol-Miro	1979	64	Male	Epigastric pain	L	1	Snare polypectomy	8
5	Murata	1980	56	Male	None	L	1.8×1.2	Snare polypectomy	9
6	Tamada	1980	46	Male	General fatigue, dysphagia, vomiting	L	Unknown	Open surgery	10
7	Arai	1981	32	Male	Dysphagia	U	Unknown	Open surgery	11
8	Liebert	1983	58	Male	Dysphagia	L	1.5	Snare polypectomy	12
9	Castellanos	1990	66	Female	Chest pain	M	2×1.5	Open surgery	13
10	Yoshida	1994	55	Male	Heartburn	M	4	Open surgery	14
11	Suwa	1996	52	Female	Dysphagia	L	2.2×2.0	Snare polypectomy	15
12	Scarpis	1998	64	Male	Epigastric pain	L	1.5	Snare polypectomy	16
13	Lee	2002	37	Male	Dysphagia	M, L	Multiple	INF α 2a+partial snare polypectomy (partial)	17
14	Yoon	2004	72	Male	Postprandial vomiting	L	5.1×2.3	Open surgery	18
15	Saers	2005	52	Female	Abdominal discomfort, dysphagia, chest pain	L	0.7	Cap-EMR	19
16	Sushil	2007	68	Male	Reflux symptoms	L	1.4×1.4	Snare polypectomy	4
17	Best	2008	63	Male	Dysphagia	U	Multiple (3.5×2.2+2×1.4)	CO ₂ laser	20
18	Seybt	2008	53	Male	Dysphagia, regurgitation	L	4	Open surgery	21
19	Binderup	2009	43	Female	Epigastric discomfort	L	0.5	Snare polypectomy	22
20	Arashiro	2010	33	Female	Reflux symptoms	L	1.9×1.5	ESD	23
21	Lee	2011	55	Male	Unknown	M	0.7	Band ligation	24
22	Xue	2012	58	Male	Dysphagia	M, L	(10×2.5×1+6×5×4)	Open surgery	25
23	Our case	2014	57	Male	None	L	1.2	Follow-up	

EMR, endoscopic mucosal resection; ESD, endoscopic submucosal dissection; INF, interferon; L, lower oesophagus; M, middle oesophagus; U, upper oesophagus.

tumour is larger and symptomatic, or when malignancy is suspected.⁴ Intraoperative endoscopy can be used to locate small lesions.²⁵ Other modalities, such as CO₂ laser excision and interferon α 2a plus partial endoscopic resection (snare polypectomy), have been used in the treatment of oesophageal lymphangiomas. Additionally, a case of endoscopic submucosal resection using band ligation was described by Lee *et al.*¹⁷

There is no evidence of oesophageal lymphangioma recurrence after complete resection in any of the reported cases. However, resection should only be performed when it is strongly indicated.

Learning points

- Oesophageal lymphangioma is an exceedingly rare submucosal tumour that is almost always found incidentally in the distal part of the oesophagus.
- Endoscopic ultrasound should be performed as part of the work up and differential diagnosis for this entity.
- Histological examination is required to confirm the diagnosis of oesophageal lymphangioma.
- Management of oesophageal lymphangiomas can be conservative in the absence of symptoms and if a definitive diagnosis has already been established; otherwise, endoscopic resection/dissection or a surgical strategy should be adopted.
- No malignant transformation or recurrence after resection of oesophageal lymphangioma has been reported.

Competing interests None declared.

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REFERENCES

- Weiss SW, Goldblum JR. *Enzinger and Weiss. Soft tissue tumors*. St. Louis: Mosby, 2008.
- Pui MH, Li ZP, Chen W, *et al.* Lymphangioma: imaging diagnosis. *Australas Radiol* 1997;41:324–8.
- Hizawa K, Aoyagi K, Kurahara K, *et al.* Gastrointestinal lymphangioma: endosonographic demonstration and endoscopic removal. *Gastrointest Endosc* 1996;43:620.
- Sushil A, Aline PC, Metin O, *et al.* Lymphangioma of the esophagus treated with endoscopic submucosal resection. *J Gastroenterol Hepatol* 2007;22:284–6.
- Watson-Williams E. Specimen: lymphangioma of the oesophagus. *Proc R Soc Med* 1934;27:1288.
- Schmidt H.W, Clagett OT, Harrison EG Jr. Benign tumors and cysts of the esophagus. *J Thorac Cardiovasc Surg* 1961;41:717–32.
- Brady PG, Milligan FD. Lymphangioma of the esophagus—diagnosis by endoscopic biopsy. *Am J Dig Dis* 1973;18:423–5.
- Armengol-Miro JR, Ramentol F, Salord J, *et al.* Lymphangioma of the oesophagus diagnosis and treatment by endoscopic polypectomy. *Endoscopy* 1979;11:185–9.
- Murata Y, Yoshida M, Hayashi T, *et al.* A case of lymphangioma of the esophagus. *Prog Dig Endosc* 1980;17:190–3.
- Tamada R, Sugimachi K, Yaita A, *et al.* Lymphangioma of the esophagus presenting symptoms of achalasia—a case report. *Jpn J Surg* 1980;10:59–62.
- Arai T, Kiuchi S. Lymphangioma of the hypopharynx and cervical esophagus. *J Jpn Bronchoesophagol Soc* 1981;32:257–62.
- Liebert CW Jr. Symptomatic lymphangioma of the esophagus with endoscopic resection. *Gastrointest Endosc* 1983;29:225–6.
- Castellanos D, Sebastian JJ, Larrad A, *et al.* Esophageal lymphangioma: case report and review of the literature. *Surgery* 1990;108:593–4.
- Yoshida Y, Okamura T, Ezaki T, *et al.* Lymphangioma of the oesophagus: a case report and review of the literature. *Thorax* 1994;49:1267–8.

- 15 Suwa T, Ozawa S, Ando N, *et al.* Case report: lymphangioma of the oesophagus endoscopically resected. *J Gastroenterol Hepatol* 1996;11:786–8.
- 16 Scarpis M, De Monti M, Pezzotta MG, *et al.* Endoscopic resection of esophageal lymphangioma incidentally discovered. *Diagn Ther Endosc* 1998;4:141–7.
- 17 Lee BI, Kim BW, Kim KM, *et al.* Esophageal lymphangiomatosis: a case report. *Gastrointest Endosc* 2002;56:589–91.
- 18 Yoon YH, Kim KH, Baek WK, *et al.* Lymphangioma of the esophagus: surgical treatment. *Ann Thorac Surg* 2004;78:e51–3.
- 19 Saers T, Parusel M, Brockmann M, *et al.* Lymphangioma of the esophagus. *Gastrointest Endosc* 2005;62:181–4.
- 20 Best SR, Coelho DH, Ahrens WA, *et al.* Laser excision of multiple esophageal lymphangiomas: a case report and review of the literature. *Auris Nasus Larynx* 2008;35:300–3.
- 21 Seybt MW, Postma GN. Giant esophageal lymphangioma. *Ear Nose Throat J* 2008;87:500.
- 22 Binderup MD, Jensen VJ, Busch S. Lymphangioma of the oesophagus. *Ugeskr Laeger* 2009;171:312–13.
- 23 Arashiro M, Satoh K, Osawa H, *et al.* Endoscopic submucosal dissection of esophageal lymphangioma: a case report with a review of the literature. *Clin J Gastroenterol* 2010;3:140–3.
- 24 Lee DG, Kim GH, Park DY, *et al.* Endoscopic submucosal resection of esophageal subepithelial lesions using band ligation. *Endoscopy* 2011;43:822–5.
- 25 Xue L, Guo WG, Hou J, *et al.* Huge lymphangiomatosis of the esophagus. *Ann Thorac Surg* 2012;93:2048–51.
- 26 Beham A. Lymphangioma. In: Fletcher CDM, Unni KK, Merten F, eds. *World Health Organization, classification of tumors. Pathology and genetics of tumors of soft tissue and bone*. Lyon, France: IARCPress, 2002: 162–3.

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