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Rare disease

Jejunal cavernous lymphangioma

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Summary

Cavernous lymphangiomas are usually identified in infants and children with the majority of lesions found around the head and neck, trunk or extremities. Tumours affecting the intra-abdominal organs are rare. The authors report a case of small bowel cavernous lymphangioma arising within the jejunum of a 34-year-old woman presenting with dyspnoea and anaemia, and review the existing literature relating to this uncommon tumour.

BACKGROUND

Cavernous lymphangiomas of the small bowel are benign lesions that often present with anaemia. As it has traditionally been difficult to investigate the ileum and jejunum, these lesions have been regarded as being rare, and often not considered in the differential diagnosis of patients with iron deficiency anaemia related to the gastrointestinal tract. With the advent of capsule endoscopy and enteroscopy, the incidence of reporting of cavernous lymphangiomas appears to be increasing. The aim of this report and literature review is to highlight this tumour within the differential for anaemia, and to identify new means of investigation that may assist in achieving the diagnosis.

CASE PRESENTATION

A 34-year-old woman was referred for investigation of dyspnoea and was found to have iron deficiency anaemia with a haemoglobin of 6 g/dl. There was no history of irregular or heavy menses and no gastrointestinal symptoms were noted. No significant medical history or

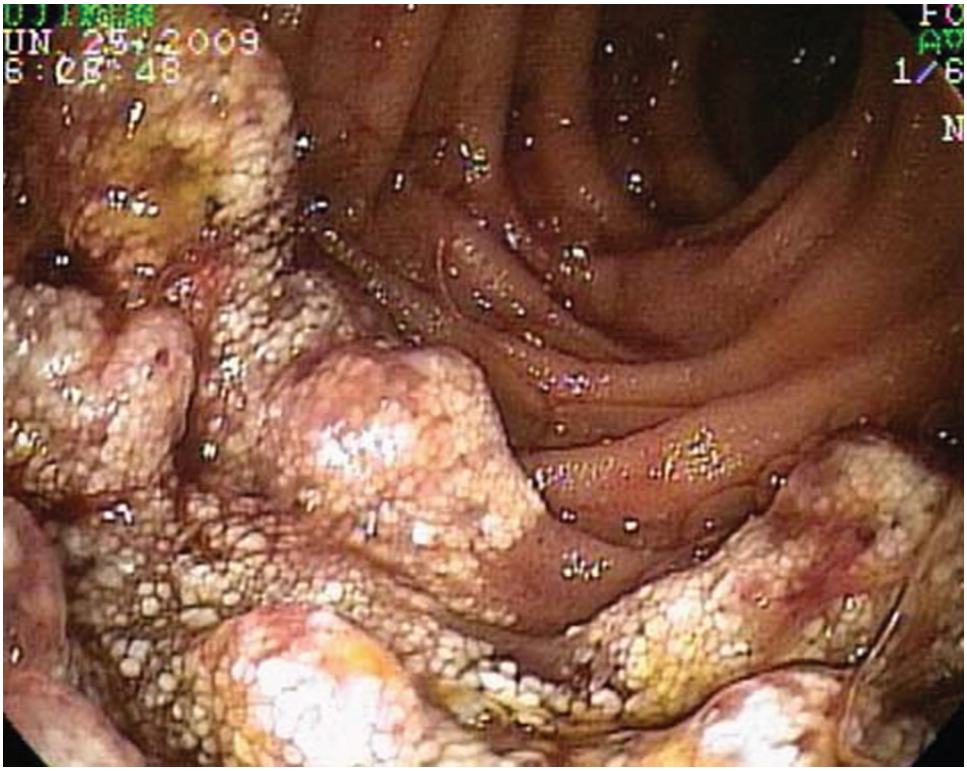

Figure 1 Enteroscopic appearance of the lymphangioma.

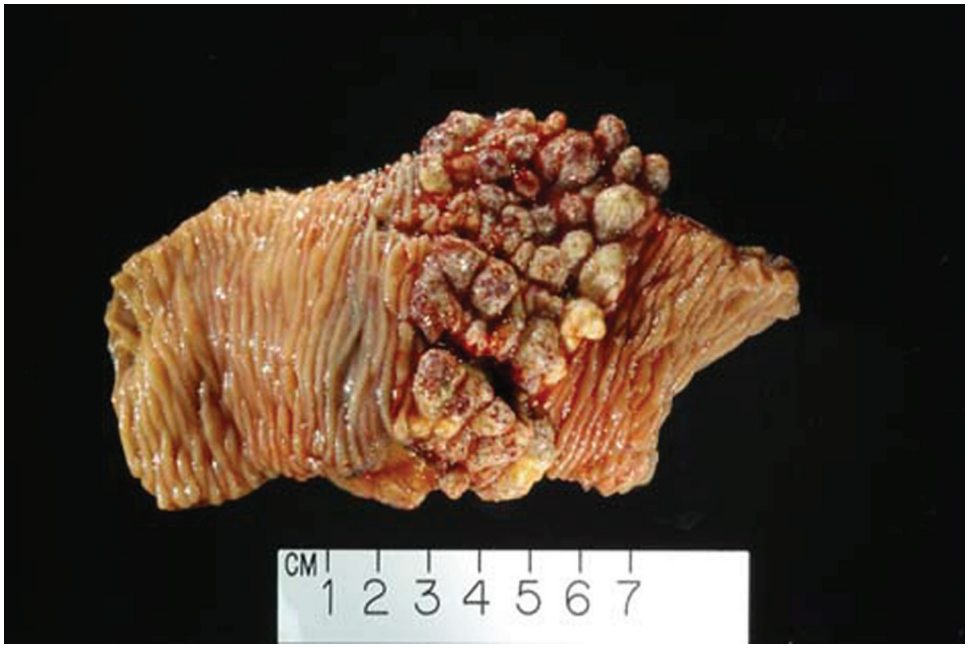

Figure 2 This gross photograph of the segmental jejunal resection illustrates the striking multinodular and polypoid appearance of the jejunal lymphangioma, along with the entirely normal appearing adjacent jejunum.

family history was noted and physical examination was unremarkable.

INVESTIGATIONS

Upper and lower endoscopies did not reveal a bleeding source. A capsule endoscopy identified a lesion within the jejunum. A CT scan identified the mass but there was no evidence of metastatic spread and the nature of the mass was uncertain. A push enteroscopy demonstrated a lobular, circumferential mass in the proximal jejunum. It was submucosal in location, occupying the whole of the circumference and had a lymphangiectatic appearance (figure 1). The appearances were classically those of a cavernous lymphangioma. The site was marked with a tattoo of India ink.

TREATMENT

An elective resection of the mass was performed. No ascites, lymphadenopathy, peritoneal disease or liver metastases were identified at laparotomy. The mass was identified by means of the tattoo placed at enteroscopy and the lesion was palpable at the marked point. A segmental resection was performed including adjacent mesentery and bowel, with an end-to-end anastomosis. An additional lesion, not identified on preoperative imaging, was found in the jejunal mesentery and was also excised.

Gross examination of the specimen revealed a polypoid, multinodular tumour measuring 5.3 × 4 × 1.5 cm with normal adjacent jejunum (figure 2). The tumour was spongy on cross-section and white fluid was expressed from cystic spaces. Histologically, the mucosa and submucosa were markedly expanded and replaced by dilated lymphatic spaces (figure 3) whereas the adjacent jejunum was normal (figure 4). The mesenteric lesion was an additional lymphangioma with fibromuscular lymphatic walls and luminal benign lymphoid aggregates (figure 5).

OUTCOME AND FOLLOW-UP

The patient's postoperative recovery was unremarkable and at follow-up she was asymptomatic.

DISCUSSION

Cavernous lymphangiomas are a rare tumour of the lymphatic system, mainly affecting the head and neck, trunk and extremities with the vast majority of tumours presenting in infancy and childhood.[1] Lymphangiomas account for 6%[2] of small bowel tumours seen in children but are a smaller percentage, approximately 1.4–2.4% in adults.[3] Within the peritoneal cavity, the majority of lymphangiomas are identified within the mesentery and retroperitoneum.[4] Lesions affecting the jejunum or ileum are rare and account for less than 1% of all lymphangiomas.[5]

The histological features of cavernous lymphangioma were first described by Gaudier and Gorse in 1913.[6] The tumour is a benign lesion consisting of single-layer endothelial-lined lymphatic spaces containing chylous or serous material.[7]

Review of the English language literature for the 50-year period from 1960 to 2009 reveals only 19 reports of small bowel lymphangiomas (table 1).[3 8–24] A total of 40 patients are reported, with a wide age range at presentation from 5 to 75 years and an equal gender distribution have been observed. Of those in which a specific anatomical small bowel location was noted, 24 of the lesions were within the jejunum and 5 in the ileum.

Hanagiri and colleagues also document an additional 33 patients from the Japanese literature between 1967 and 1990. In addition to the 19 English literature reports, 8 were from the Far East, including 5 Japanese and 3 Taiwanese manuscripts, suggesting that there may be a Japanese/Taiwanese predisposition to small bowel lymphangiomas. In the Japanese language literature, although the age distribution was comparable, there were twice as many males

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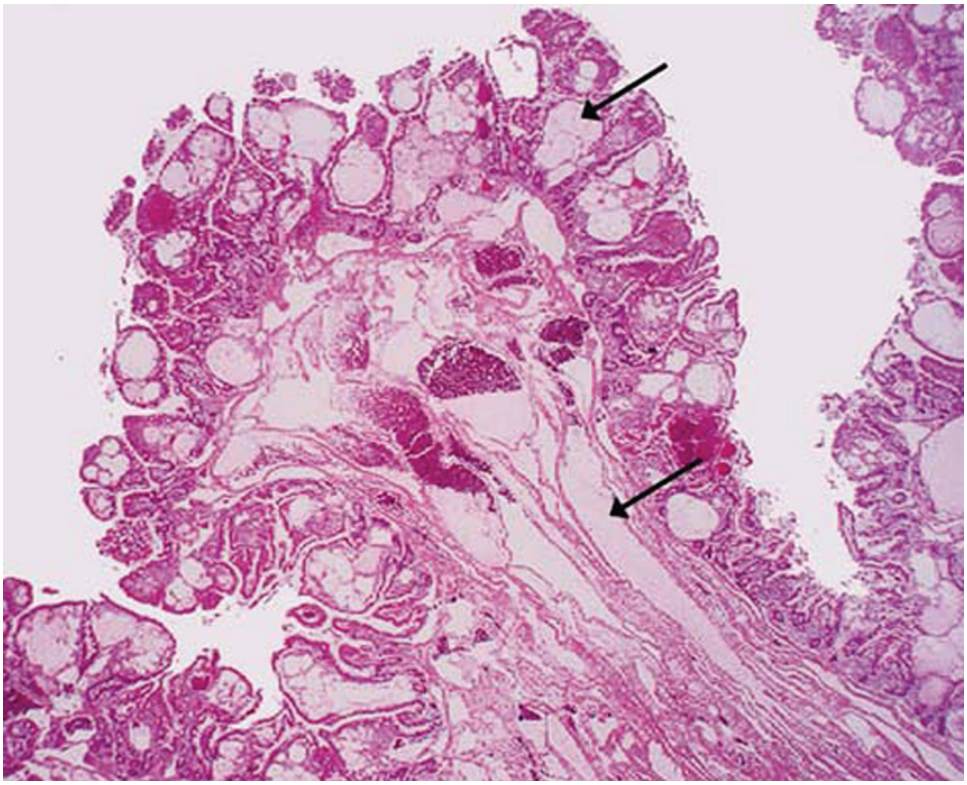

Figure 3 The mucosa and submucosa are expanded and replaced by innumerable dilated lymphatics (arrows).

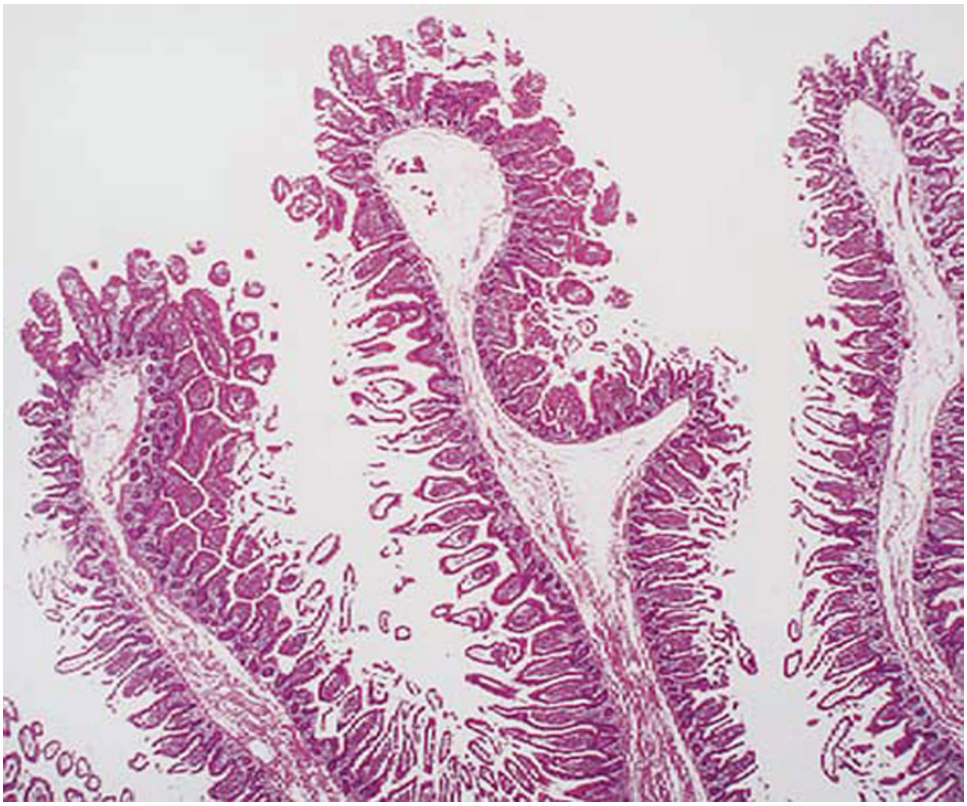

Figure 4 The jejunum adjacent to the lymphangioma was normal histologically, as illustrated here with intact villous architecture, mucosa and submucosa, lacking dilated lymphatic spaces. This feature excludes diffused lymphangiomatosis from the differential.

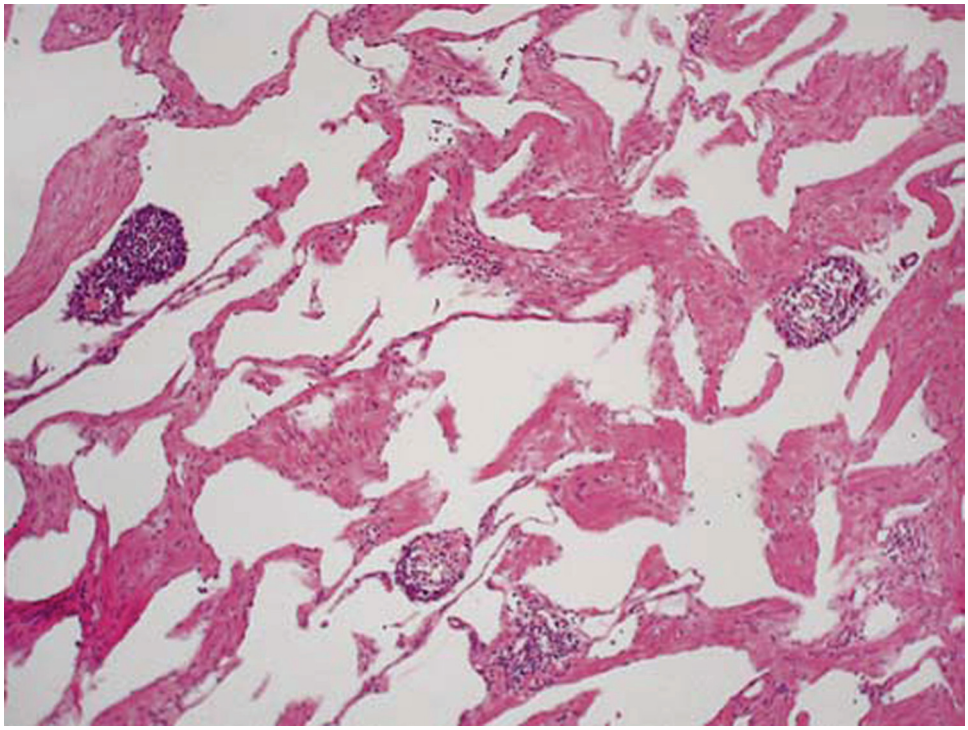

Figure 5 In addition to the mass forming lymphangioma occupying the wall of the jejunum, an additional focal mesenteric lymphangioma was identified in the resection specimen. The fibromuscular walls making up the lymphatic vessels and luminal benign lymphoid aggregates are illustrated.

Table 1 English literature summary of jejunal and ileal lymphangioma case reports

Author	Year	No	Gender	Age	Presentation	Location	Other lesions	Treatment
Shyung	2009	2	M	32	Anaemia	NS	No	DBE
			M	43	Incidental	NS	No	NS
Feng	2009	1	M	69	Melaena	Jejunum	No	DBE
Yeh	2009	1	NS	NS	GI bleeding	Ileum	No	Resection
Crook	2008	1	M	33	GI bleeding	Jejunun	No	Resection
Huang	2008	1	M	51	Melaena	Jejunum	No	Resection
Norris	2008	1	F	23	Ovarian mass	Jejunum	Yes	Resection
Hsu	2007	1	F	75	Anaemia	Jejunum	No	Resection
Pickhardt	2007	1	F	55	Asymptomatic	Ileum	No	Resection
Oshita	2005	1	M	5	Intussusception	Jejunum	NS	Resection
Honda	2003	1	F	31	Anaemia	Jejunum	No	Resection
Seki	1998	1	F	54	Abdominal mass	Jejunum	Omentum (main)	Resection
Uncu	1997	2	M	43	Abdominal mass	Ileum	No	Resection
			M	19	Incidental	Ileum	No	Resection
Hanagiri	1992	1	M	53	Obstruction	NS	No	Resection
Shigematsu	1988	3	M	55	Anaemia/melaena	Jejunum (2) + ileum (1)	No	Resection
			F	65	Incidental	Jejunum (7)	Duodenum	Resection
			M	46	Abdominal pain	Jejunum (2)	No	Resection
Colizza	1981	1	F	58	Melaena	Jejunum	Stomach	Resection
Cohen	1971	1	NS	NS	NS	Jejunum	NS	NS
Schmutzer	1964	3	NS	NS	Incidental	Jejunum	No	Resection
			NS	NS	Incidental	Ileum	No	Resection
			NS	NS	Incidental	NS	No	Resection
Good	1962	16	NS	NS	NS	NS	NS	NS
Ralston	1961	1	M	58	Anaemia	Jejunum	No	Resection

DBE, double-balloon enteroscopy; GI, gastrointestinal; NS, not stated.

as females with an equal distribution between ileum and jejunum.

The number of lymphangiomas reported has increased in recent literature, with 10 of the 19 papers published in the past decade. This is likely due to improvements in diagnostic imaging with the advent of capsule endoscopy and double-balloon enteroscopy rather than a true increase in the prevalence of the tumour.

The standard management of lymphangiomas until recently has been through surgical resection. However,

with the advent of double-balloon enteroscopy, this modality may be able to treat small tumours and this indeed has been accomplished in two cases.[23 24]

Learning points

- Cavernous lymphangiomas are rare tumours of the lymphatic system, mainly affecting the head and neck, trunk and extremities usually presenting in infancy and childhood.
- Lesions affecting the small bowel are uncommon but appear to be on the increase probably due to improvements in diagnostic modalities available.
- The standard management of lymphangiomas until recently has been through surgical resection, however, the development of double-balloon enteroscopy has been successfully used to treat small tumours and may become the standard of care in the future.

Competing interests None.

Patient consent Obtained.

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