

〈症 例 報 告〉

Cavernous Lymphangioma of the Intestines : Report of Two Cases

Masayuki Shintaku, M.D.

Department of Pathology, Osaka Red Cross Hospital

Tsutomu Hachiya, M.D.

Department of Health Care, Osaka Red Cross Hospital

Sadayuki Matsuoka, M.D.

Department of Surgery, Osaka Red Cross Hospital

Hitoya Ohta, M.D.

Department of Laboratories, Osaka Red Cross Hospital

ABSTRACT

Two cases of cavernous lymphangioma arising in the intestinal wall are reported. The first case, a 74-year-old man, had an intramural cavernous lymphangioma in the duodenum detected on computerized tomographic examination and magnetic resonance imaging. The second case, a 59-year-old woman, had a large polypoid cavernous lymphangioma in the ascending colon detected on barium enema examination. No clinical symptoms referable to lymphangiomas were found in both cases. Radiological examination is thought to be a useful means of detecting lymphangiomas of the intestines, although preoperative diagnosis of the exact nature of these lesions can be difficult because of their extreme rarity.

Key words : lymphangioma, duodenum, colon

INTRODUCTION

Lymphangiomas arising in the abdominal organs are rare, and the majority of them arises in the mesentery, omentum, mesocolon, and retroperitoneum¹⁾. Those arising in the wall of the gastrointestinal tract are even more rare, and according to Davis et al.²⁾ only 39 cases of intestinal lymphangioma have been reported until 1987. Although the diagnosis of lymphangiomas is usually established by pathological examination, the preoperative radiological examination occasionally enables the precise delineation of the lesions and suggests correct diagnosis.

We present here the radiological and pathological features of two adult cases of abdominal cavernous lymphangioma arising from unusual locations. In the first case, the tumor was found within the duodenal wall on computerized tomographic examination (CT) and magnetic resonance imaging (MRI). In the second case, the tumor had grown to a large pedunculated polyp of the ascending colon and was detected on barium enema examination.

CASE REPORT

Case 1 : The patient was a 74-year-old man, who presented with lower abdominal pain and

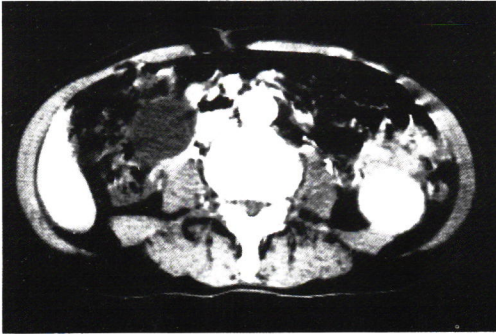


Fig. 1 Case 1. CT scan with contrast enhancement of the abdomen demonstrated a round, non-enhanced cystic mass outside of the duodenal loop.

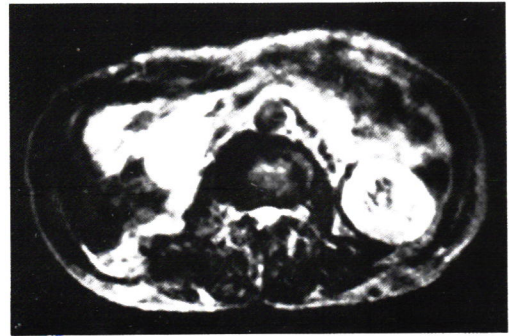
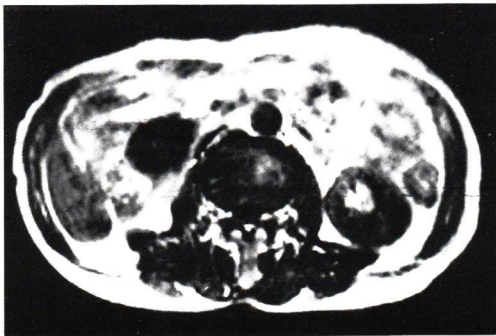


Fig. 2 & 3 Case 1. MRI of the duodenal cystic lesion showed a low signal intensity on T1-weighted image (Fig. 2) and a very high signal intensity on T2-weighted image (Fig. 3).



Fig. 4 Case 1. Gross appearance of the duodenal tumor at autopsy. The tumor showed a multilocular cystic appearance with a smooth transparent surface.

lumbago. Thorough examination revealed adenocarcinoma of the lung with metastasis to the vertebral column and retroperitoneum. CT of the abdomen disclosed a low density mass without contrast enhancement, measuring about 4 cm in diameter, just lateral to the duodenal loop (Fig. 1). On MRI, the lesion showed a low signal on T1-weighted image (600/13) (Fig. 2) and a very high signal on T2-weighted image (2500/80) (Fig. 3). Ultrasound-guided puncture of this cystic lesion yielded clear, serous fluid. The patient developed paraplegia, then died of carcinomatous cachexia and bronchopneumonia. Autopsy revealed extensive metastasis of pulmonary adenocarcinoma to various organs.

In the lateral wall of the second portion of the duodenum, a multilocular cystic tumor, measuring 4.5×3.5×3.5 cm was found outside of the duodenal muscle layer (Fig. 4). It had a

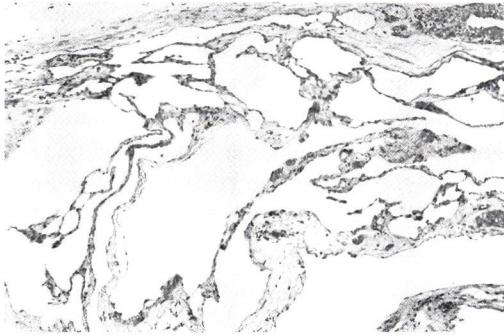


Fig. 5 Case 1. The tumor consisted of irregular lymphatic spaces lined by flat endothelial cells. The spaces were separated by thin fibrous septa. (Hematoxylin - Eosin stain, $\times 25$)

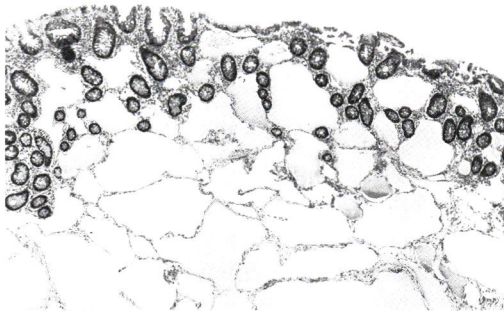


Fig. 7 Case 2. In the mucosal portion of the tumor, many intestinal crypts separated the dilated lymphatic spaces. (Hematoxylin-Eosin stain, $\times 10$)

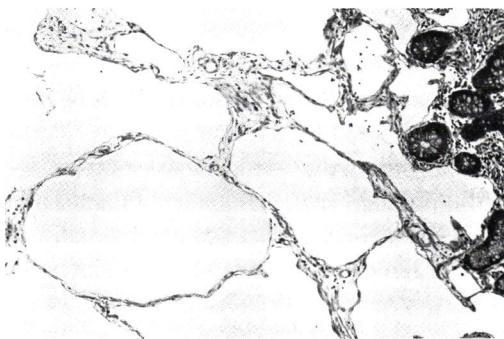


Fig. 8 Case 2. Lymphatic spaces with irregular shapes and sizes were lined by flat endothelial cells and contained pale eosinophilic fluid. (Hematoxylin-Eosin stain, $\times 25$)

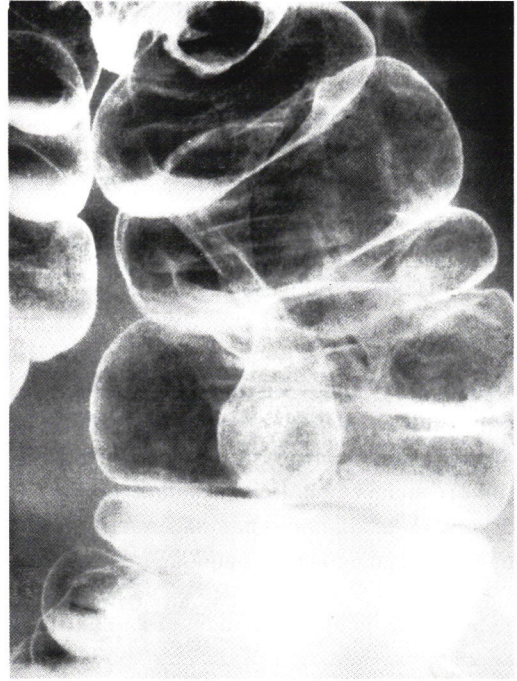


Fig. 6 Case 2. Barium enema examination showed a large pedunculated polypoid lesion in the ascending colon. (P-A view)

smooth transparent surface and contained clear serous fluid. On cut section, the tumor was comprised of aggregates of tiny irregular spaces, each measuring a few millimeters in diameter, which were separated from each other by thin fibrous septa.

Histopathologically, the tumor consisted of lymphatic spaces of irregular shapes and sizes separated by fibrous septa occasionally containing small amounts of smooth muscle fibers (Fig. 5). Each space was lined by a single layer of bland-appearing, flat endothelial cells. Mild, focal lymphocytic infiltration was noted in the septa.

Case 2: The patient was a 59-year-old woman, who presented with anal bleeding that persisted for two months. Barium enema and endoscopic examination revealed a large, circumferentially protruding lesion in the sigmoid colon, and biopsy specimens taken from this lesion were diagnosed as

adenocarcinoma. While the fiberoptic could not be passed beyond this lesion, barium enema study depicted a large pedunculated polyp having a smooth surface and a long stalk in the ascending colon (Fig. 6). Sigmoidectomy and resection of the polyp by means of colotomy was performed, and the patient recovered uneventfully. Hypoalbuminemia and other symptoms and signs suggestive of a protein-losing enteropathy were not found.

The resected polyp measured 25×16×17 mm and had a yellowish, transparent smooth surface. It was connected to the colonic mucosa with a thin stalk approximately 24 mm long. On cut section, the polyp, which showed a honeycomb appearance, consisted of aggregates of microcysts, each measuring less than 2 mm in diameter and containing clear fluid. The central area of the polyp was slightly hard and whitish.

Histopathologically a large number of irregularly shaped lymphatic spaces filled with pale eosinophilic fluid were found extending from the lamina propria to the submucosal region of the polyp (Fig. 7). Each of these spaces was lined by flat endothelial cells showing no cytological atypism and separated by a thin fibrous septum (Fig. 8). In the intramucosal portion of the tumor, each lymphatic space was separated by intervening mucosal tissue containing intestinal crypts. Variable amounts of smooth muscle fibers, some of which were in continuity with the muscularis mucosae around the lesion, were found in the septa.

DISCUSSION

The two cases presented here showed almost typical gross and microscopic features of cavernous lymphangioma. In the second case, histopathological appearance of the tumor was somewhat unusual in that the tumor involved the lamina propria extensively and each dilated lymphatic channel was separated by mucosal tissue containing intestinal crypts. Lymphangiomas of the intestines are usually located in the submucosa with the overlying mucosa remaining intact³⁾⁻⁵⁾. The diagnostic possibility of intestinal lymphangiectasia^{6),7)} was considered in this case, but similar lymphangiectatic lesions were not found either in the stalk of the polyp or in the resected specimen of the sigmoid colon, and there were no other clinical or radiological features suggesting the presence of intestinal lymphangiectasia.

Most of the examples of lymphangioma of the abdominal cavity arise in the mesentery^{1),8)}, and those arising within the intestinal wall or presenting as intraluminal polypoid lesions are very rare^{4),5)}. Lymphangiomas are in general regarded as a kind of hamartomatous or malformative lesions, and many of them are detected during childhood¹⁾. However, intra-abdominal lymphangiomas found in adults are not uncommon. All reported cases of lymphangioma of the large intestine which were reviewed by Berardi⁵⁾ had been found in adults. These are occasionally asymptomatic and found incidentally at autopsy or surgical exploration for other reasons⁷⁾. Because of its extreme rarity, specific preoperative radiological diagnosis of lymphangiomas of the intestines is very difficult, even if the presence of lesions can be detected. According to a review by Berardi⁵⁾, none of the cases of lymphangioma of the large intestine was diagnosed preoperatively. Elucidation of the exact nature of the lesions must usually await pathological examination of the resected specimens.

In our first case, the cystic nature of the duodenal lesion was clearly demonstrated by CT, and the MRI indicated that the content of the cyst was not viscid. Clear, serous fluid was obtained by puncture of the lesion. From these findings, duplication cyst of the duodenum and microcystic adenoma of the pancreatic head were considered the most likely diagnostic possibilities. The extreme rarity of lymphangioma arising in the duodenum^{2),9)} prevented the

correct antemortem diagnosis. According to Davis et al.²⁾ only eight of 39 reported cases involving lymphangioma of the intestines arose in the duodenum. Lymphangioma should be included in the list of radiological differential diagnosis of intramural tumors of the intestines, although its incidence is extremely low.

In the second case, polypoid lesion of the ascending colon was considered radiographically most likely to be an epithelial (adenomatous) polyp. In a series of lymphangiomata of the large intestine reviewed by Berardi⁵⁾, most of the cases were diagnosed as adenomatous polyp or adenocarcinoma preoperatively. However, the mucosal surface of polypoid lesions is unusually smooth, which, as noted earlier by Arnett and Friedman³⁾, is rather suggestive of submucosal mesenchymal neoplasm, especially lipoma.

It is generally agreed that lymphangiomata are hamartomatous or malformative lesions which arise from sequestrations of lymphatic tissue that failed to communicate normally with the lymphatic system^{1),7)}. Most of them are considered present at birth¹⁾. However, in cases of lymphangioma appearing late in adulthood, the possibility that the lesions have developed secondarily from acquired lymphangiectasia resulting from local disturbance of lymphatic circulation is not excluded¹⁰⁾. In our first case, extensive metastasis of pulmonary adenocarcinoma was found in the vertebral column and the surrounding retroperitoneal tissue. It is likely that the local disturbance of lymphatic circulation caused by metastatic carcinoma triggered lymph stasis and secondary lymphangiectasia which in turn caused the formation of a lymphangioma in that very unusual location.

REFERENCES

1. Enzinger FM & Weiss SW : Soft tissue tumors, 3rd ed., Mosby, St. Louis, 1994, p. 679-700.
2. Davis M, Fenoglio-Preiser CM, Haque AK : Cavernous lymphangioma of the duodenum. Case report and review of the literature. *Gastrointest Radiol* 12 : 10-12, 1987.
3. Arnett NL & Friedman PS : Lymphangioma of the colon : Roentgen aspects. A case report. *Radiology* 67 : 882-885, 1956.
4. Greene EI, Kirshen MM, Greene JM : Lymphangioma of the transverse colon. *Amer J Surg* 103 : 723-726, 1962.
5. Berardi RS : Lymphangioma of the large intestine. Report of a case and review of the literature. *Dis Colon Rectum* 17 : 265-272, 1974.
6. Ivey K, DenBesten L, et al. : Lymphangiectasia of the colon with protein loss and malabsorption. *Gastroenterol* 57 : 709-714, 1969.
7. Fenoglio-Preiser CM, Pascal RR, Perzin KH : Tumors of the intestines. *Atlas of Tumor Pathology, Second series, Fasc. 27*. Armed Forces Institute of Pathology, Washington, D. C., 1990, p. 494-500.
8. Daniel S, Lazarevic B, Attia A : Lymphangioma of the mesentery of the jejunum. Report of a case and a brief review of the literature. *Amer J Gastroenterol* 78 : 726-729, 1983.
9. Elliott RL, Williams RD, et al. : Lymphangioma of the duodenum. Case report with light and electron microscopic observation. *Ann Surg* 163 : 86-92, 1966.
10. Fisher I & Orkin M : Acquired lymphangioma (lymphangiectasis). Report of a case. *Arch Dermat* 101 : 230-234, 1970.