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CASE REPORT

Cystic Lymphangioma of the Small-Bowel Mesentery

Case Report and a Review of the Literature

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Cystic lymphangioma of the small-bowel mesentery is a rare manifestation of an intraabdominal tumor in elderly patients. We present a case of a small-bowel mesentery lymphangioma, causing fever and chills and present clinical and pathologic features. Furthermore, etiology and differential diagnosis of this tumor are discussed. (Pathology Oncology Research Vol 6, No 2, 146–148, 2000)

Keywords: lymphangioma, small-bowel mesentery, infection, etiology, differential diagnosis

Introduction

Lymphangiomas are rare tumors. As many as 90% may manifest before the age of three[2] and the sex ratio is roughly equal in childhood.[11] In young patients lymphangiomas are preferentially located in head, neck and axilla, but they also occur sporadically in various parenchymal organs (e.g. spleen, liver, bones), sometimes as a diffuse or multifocal disease (lymphangiomatosis). During adulthood, they mostly appear as superficial cutaneous lymphangioma or as intraabdominal lymphangiomas. There is a male to female ratio for intraabdominal lymphangiomas of 3:1.[16] Herein, we present a case of a cystic lymphangioma of the small-bowel mesentery and discuss etiology and differential diagnosis.

Case report

A 61 year old man was refered to the hospital at the end of 1997 with abdominal pain of the lower left abdomen and elevated temperatures. Ultrasound raised suspicion of a peridiverticulitis of the sigma. Barium enema, however, did not confirm the diagnosis. In view of the fact that the clinical symptoms resolved further diagnostic intervention (CT) was not performed. Six months later the patient was readmitted with abdominal pain, elevated temperature, and leucocytosis . He further had chills and was confused. Ultrasound showed the same picture as 6 month before. CT (*Figure 1*) revealed a large tumourous process in the pelvis surrounding some loops of the small intestine. X-ray findings on contrast study of the small bowel showed an ileum loop draped around a mesenteric mass with compression of the bowel wall (*Figure 2*). Under antibiotic therapy and total parenteral nutrition the symptoms resolved and 6 days later the patient underwent surgery. 30 cm of the terminal ileum with the adjunct tumorous mass in the mesentery was resected. On the 12 th postoperative day the patient was discharged in a good physical condition with no remaining clinical symptoms.

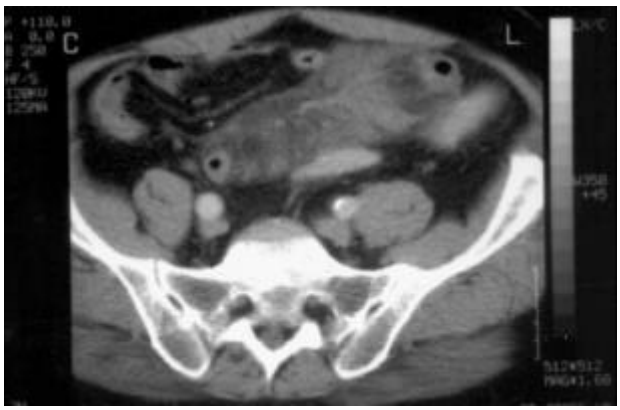

Figure 1. CT showing a large tumourous process in the pelvis

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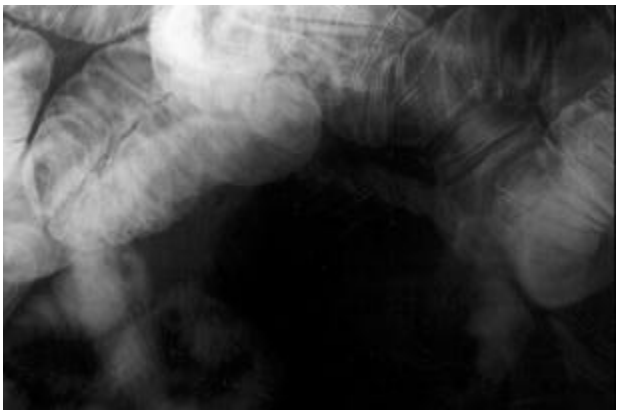

Figure 2. X-ray findings on contrast study of the small bowel showing an ileum loop draped around a mesenteric mass

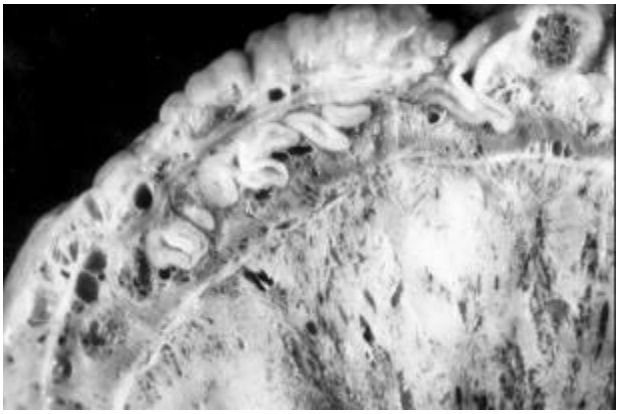

Figure 3. Cross- sections showing a multicystic tumour involving all parts of the bowel wall

Pathology

The resected segment of ileum (30 cm) showed a large, well-circumscribed, polycystic mass in the mesentery (12x9x7 cm). The cross-sections (*Figure 3*) showed a multicystic tumour containing clear fluid and involving all parts of the bowel wall. Microscopically, the histological sections showed variable sized, markedly dilated lymphatic channels in the mesentery and all parts of the bowel wall lined by flat endothelial cells (*Figure 4*), that were positive in immunohistochemistry studies for Factor VIII related antigen and Ulex europaeus. The same cells showed no reaction for cytokeratin. Some lymphatic spaces contained proteinaceous fluid, secondary intralesional hemorrhage was also present. The wall of the spaces was built up of fibroconnective tissue accompanied by aggregates of lymphoid tissue as well as normal arteries and veins. Moreover, fascicles of smooth muscle as well as collagen bundles could be seen. The final diagnosis was cystic lymphangioma.

Discussion

Although lymphangiomas are benign lesions, they may cause significant mortality because of their large sizes, critical locations and the possibility of becoming secondarily infected. Almost all lesions require surgical treatment. The extend of the procedure should be dictated by the location and the desire to achieve a reasonable cosmetic result especially for lymphangiomas of the head and neck. However, incomplete removal can result in recurrence even after many years.[15]

Intraabdominal lymphangiomas are rare tumours, accounting for approximately 1 per 100.000 hospital admissions.[10] They occur in the mesentery of the small and large bowel but also at retroperitoneal sites. The clinical symptoms range from chronic to acute abdominal pain sometimes due to perforation,[10] obstruction[3,9,12] or anaemia due to hemorrhage.[1]

The etiology of lymphangiomas is still a matter of discussion. A well established theory suggests,[6] that lymphangiomas arise from sequestrations of lymphatic tissue during embryologic development. On the other hand Godart[7] postulated that premature lymphatics appear as mesenchymal slits, which coalesce and normally communicate with the venous system. 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 the lymphatic sacs occur.

In adults, however, sequestration of lymphatic tissue is most likely secondary to inflammatory processes or surgical or radiation therapy. The theory of secondary development would explain lymphangioma reports of patients over 60 years, who had negative radiologic findings a few years before the disease was diagnosed.[1,4,8,14]

In 1828, Radenbacker first described a cystic lymphangioma.[5] Traditionally, lymphangiomas have been divided into three groups.[13] Simple capillary, cavernous and cystic

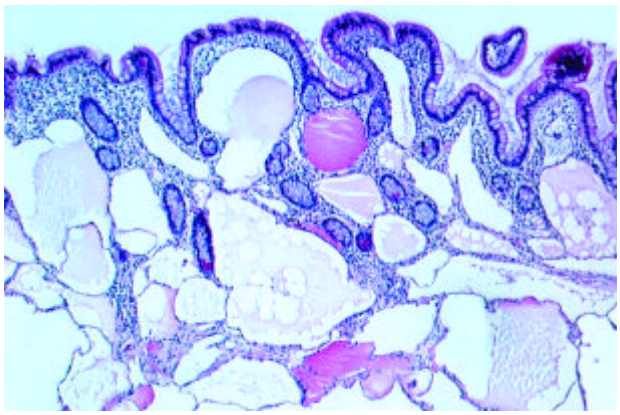

Figure 4. Low power view of the cystic lymphangioma

lymphangiomas. The capillary lymphangioma is composed of small thin-walled lymphatics, whereas the cavernous lymphangioma consits of larger lymphatics with adventitial coats. The cystic lymphangioma consists of large macroscopic lymphatic spaces that possess investitures of collagen and smooth muscle. In most cases the diagnosis is straight forward for intraabdominal lymphangiomas. The differential diagnosis includes cavernous haemangiomas, when they show secondary hemorrhage, mesotheliomas, and tumors of the pancreas. The diagnosis of a lymphangioma over a haemangioma can be favoured when there are lymphoid aggregates in the stroma and more irregular lumina with widely spaced nuclei, as shown in our case. One should always be aware of the cystic form of mesotheliomas as well as serous cystadenoma (synonym: microcystic adenoma, glycogen-rich cystadenoma) and carcinoma of the pancreas. Although lymphangioma is a benign lesion, it should be remembered that after irradiation transformation into a lymphangiosarcoma is possible.

In conclusion, intraabdominal lymphangiomas are rare benign tumors that occur in the mesentery of the large and small bowel. Symptoms of these tumors may mimic a wide spectrum of diseases; in our patient ultrasound raised initially suspicion of a peridivertikulitis. In the case presented here the etiology might be due to infection, as the patient presented twice with abdominal pain and fever. Complete excision was the treatment of choice.

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