Cavernous lymphangioma of the Small-Bowel Mesentery: A case report

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OBJECTIVES

Lymphangioma (LA) is an uncommon malformation of lymphatic system, which shows benign proliferation of lymph vessels in the mesentery and all parts of the bowel wall, the latter with the characteristics of submucosal tumors covered with normal mucosa. The incidence of LA in the gastrointestinal tract is low, and most cases are solitary. Herein, we present a case of a LA of the small-bowel mesentery and discuss differential diagnosis.

METHODS

A 16-year-old young-man with abdominal pain of the lower abdomen and constipation was admitted to our hospital. He had no significant past medical history. Physical examination revealed no remarkable abnormality in the abdomen. Laboratory rests (samples) showed hypochromic microcytic anemia. Tumor marker levels (CEA, CA19-9) were within normal ranges. X-ray findings on contrast study of the small bowel showed an ileum loop dropped around a mesenteric mass. The patient underwent surgery.

RESULTS

49cm of the terminal ileum with the adjunct mesentery was resected. The cross-sections of the resected segment showed a mass of 5 cm in the mesentery, involving all parts of the bowel wall. There are several protruding ileal mucosal lesions covered with normal mucosa, raging 3-5 mm in diameter. Microscopically the sections showed markedly dilatated lymphatic channels, lined by a single very attenuated layer of flattened endothelial cells with no atypia and contained small amounts of proteinaceous fluid, with few erythrocytes. 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. Endothelial cells were positive for all vascular markers used (FVIII, CD31, CD34), negative for cytokeratin, EMA, calretinin, HBME. Spindle cells were found arranged in poorly delineated fascicles, positive for vimentin, desmin, alpha smooth muscle actin, negative for PGR, ER, SI00, HMB45. The final diagnosis was cavernous lymphangioma.

Fig.1 Section shows markedly dilatated lymphatic channels.
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Fig.2: Innumerable ramifying empty spaces

Fig.3: Lymphatic channels lined by a single very attenuated layer of flattened endothelial cells with no atypia

Fig.4: Fascicles of smooth muscle as well as collagen bundles could be seen

Fig.5: Endothelial cells were positive for vascular marker CD-31
CONCLUSIONS

In this case there is not prominent synchronous involvement of viscera, bones and internal soft tissues of the trunk and pelvis. The differential diagnosis include cavernous haemangioma, there is no reliable endothelial markers that can distinguish blood vessel from lymphatic endothelium, however there is an important histologic difference as the former is characterized by large thick-walled irregular spaces with extensive dissection of collagen bundles. A conservative surgical treatment with segmental intestinal resection and termino-terminal anastomosis was curative.

Fig.6: Endothelial cells were positive for vascular marker CD-34

References