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**Peritoneal cavernous hemangiomatosis: A case report**

Fu LY *et al.*Peritoneal cavernous hemangiomatosis

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**Abstract**

***BACKGROUND***

Cavernous hemangiomatosis in the liver and spleen has been reported, but it occurs less commonly in the peritoneum. Here we report a case of peritoneal cavernous hemangiomatosis and share some valuable information about this disease.

***CASE SUMMARY***

A 57-year-old Chinese man had a huge abdominal mass with abdominal distention and a significant reduction of food consumption. An enhanced abdominal and pelvic computed tomography and positron emission tomography–computed tomography revealed multiple cystic masses on the peritoneum, greater omentum, small intestinal mesentery and the surface of the spleen, and a high maximum standardized uptake value of the largest cystic lesion. Exploratory laparotomy was performed, and multiple cystic masses were found on the surface of the peritoneum, greater omentum, mesentery of the small intestine, and surface of the liver and spleen. Dark red bloody cystic fluid was present in the cystic tumor. Pathological examination showed that in the stromal components, the irregular vascular wall was thin. The vessel lumen was interlinked, and the lumen was lined with squamous endothelium. According to the intraoperative findings and pathologic results, the patient was diagnosed with peritoneal cavernous hemangiomatosis.

***CONCLUSION***

The possibility of peritoneal cavernous hemangiomatosis should be considered when multiple cystic masses are found in the abdominal cavity by preoperative examination.

**Key words:** Peritoneum; Hemangiomatosis; Cavernous hemangiomatosis; Cystic lesion; Case report

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**Core tip:** Cavernous hemangiomatosis in the liver and spleen has been reported, but it rarely occurs diffusely in the abdominal cavity. In 2011, Ribback *et al* reported a case of nodular hemangiomatosis of pleura and peritoneum. We report here a case of peritoneal cavernous hemangiomatosis, along with its pathological type and positron emission tomography–computed tomography findings, for the first time. This case may help us to better understand this disease with regard to clinical manifestations and laboratory examination, imaging and pathologic results.

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**INTRODUCTION**

Hemangiomatosis can occur simultaneously in the thoracic and abdominal cavities[1]. Peritoneal cavernous hemangiomatosis occurs only in the abdominal cavity. Cystic lesions are widely distributed in the peritoneum, and dark red bloody cystic fluid is found in the lesions. The diagnosis of peritoneal cavernous hemangiomatosis mainly depends on pathological examination. Herein, we report a case of peritoneal cavernous hemangiomatosis and provide some valuable information about this disease.

**CASE PRESENTATION**

***Chief complaints***

A 57-year-old man was admitted to our hospital for evaluation of a huge abdominal mass causing abdominal distension and a significant reduction of food consumption for nearly 2 mo.

***History of past illness***

His past history was unremarkable.

***Personal and family history***

His family history was unremarkable.

***Physical examination upon admission***

Physical examination revealed a huge, tough mass. The upper boundary of the mass was below the right costal margin. The lower boundary, which was in the pelvic cavity, was hard to touch. The surface of the mass was smooth. The mass was unmovable, and there was no tenderness associated with the mass.

***Laboratory examinations***

Routine blood parameters were within the normal range. Tumor marker measurement results were as follows: carcinoembryonic antigen, 0.00 ng/mL; alpha-fetoprotein, 4.10 ng/mL; CA19-9, 9.80 U/mL; and CA72-4, 1.19 U/mL. An enhanced abdominal and pelvic computed tomography scan revealed a right mid-lower abdominal cystic lesion of 35.6 cm × 14.3 cm × 9.6 cm compressing the intestine and right ureter. Multiple small cystic masses appeared on the peritoneum, greater omentum, small intestinal mesentery, and the surface of the spleen (Figure 1A). Perihepatic effusion was also present. There was a huge mass in the abdominal cavity and the gastrointestinal tract structure was displaced. In order to exclude the extensive metastasis of gastrointestinal tumors, the patient underwent positron emission tomography–computed tomography. The examination revealed a high maximum standardized uptake value (SUVmax) of the largest cystic lesion. The SUVmax of the lesion was 2.7. Mild radioactivity uptake was seen in the multiple cystic lesions on the peritoneum, omentum, small bowel mesentery, and surface of the liver and spleen. Possible preoperative diagnosis was peritoneal metastasis of appendix mucinous tumor.

 Explorative laparotomy revealed multiple dark red masses on the small intestinal mesentery, peritoneum, and greater omentum, and surface of the liver and spleen. The diameter of the masses ranged from 2 cm to 5 cm. Dark red bloody cystic fluid was seen when the thin wall was incised. A huge soft tumor was found in the right abdominal cavity. The huge cystic lesion was tightly adhered to the small intestine and ascending colon, making it difficult to be separated from the organs. Dark red bloody cystic fluid was present in the mass. An incisional biopsy of the cystic wall and some other smaller cystic lesions was obtained (Figure 1B). Intraoperative frozen section showed that the inner cystic wall was lined with squamous endothelium without atypia. However, the high SUVmax and the wide distribution of the masses indicated the possibility of malignancy. Although the sensitivity and specificity of intraoperative frozen pathological examination was very high, we used treatments for malignant tumors considering that the malignant possibility could not be completely excluded after a consultation with the pathologist. We then performed complete aspiration of the seroperitoneum and successively embrocated iodine and carbolic acid on the inner wall of the largest mass. The patient underwent hyperthermic perfusion with 4000 mL normal saline at 43 ºC for 1 hr immediately after surgery.

Pathological examination showed that within the stromal components, the irregular vascular wall was thin. The vessel lumen was interlinked, and the lumen was lined with squamous endothelium without heterogeneity. Hemorrhage and hemosiderin were present in the stromal components. Immunohistochemical staining of the endothelial cells showed positivity for CD31 and ERG and negativity for D2-40 (Figure 1C). The Ki-67 value was 2%. The diagnosis was peritoneal cavernous hemangiomatosis according to the intraoperative findings and pathologic results.

**FINAL DIAGNOSIS**

According to the pathological examination, the final diagnosis was peritoneal cavernous hemangiomatosis.

**TREATMENT**

Explorative laparotomy was performed because of multiple abdominal masses causing abdominal distension and the diagnosis was not clear. After surgery, the patient underwent hyperthermic perfusion immediately because the possibility of a malignant disease cannot be completely excluded.

**OUTCOME AND FOLLOW-UP**

The patient returned to Shanxi Province after operation and did not go to our hospital for examination. He underwent an abdominal ultrasound examination in a local hospital 2 wk previously. The result showed that multiple cystic masses still existed in the abdominal cavity, but the patient had no discomfort.

**DISCUSSION**

Here, we report a case of peritoneal cavernous hemangiomatosis. Cavernous hemangiomatosis can occur in the parenchymal viscera such as liver[2] or spleen[3]. In 2011, Ribback *et al*[1] reported a case of nodular hemangiomatosis of the pleura and peritoneum; however, the advanced pathological type of the hemangiomatosis was not described.

According to the pathology and immunohistochemistry results, the lesion in the present case was composed of vascular components without lymphatic components. Therefore, we excluded the diagnosis of lymphangioma[4] and other lymphangiogenic tumors. We also excluded the diagnosis of epithelioid hemangioma since the vascular lumen was lined with squamous endothelium without lymphocytes, eosinophils, or the formation of lymphoid follicles, and imaging examination did not show any abnormal soft tissue mass and bone destruction[5,6]. Platelet count was not decreased in this patient and no spindle tumor cells or fissured lacuna were found in pathological examination, allowing us to further exclude the diagnosis of kaposiform hemangioendothelioma[7,8]. The diagnosis of multicystic mesothelioma was also excluded as the mass contained dark red cystic fluid and the inner cystic wall was lined with flat endothelial cells instead of cubic mesothelial cells[9]. Pathological examination showed that the lumen was lined with squamous endothelium without heterogeneity, and immunohistochemical examination showed that the Ki-67 value of the endothelial cells was 2%. We excluded the possibility of malignant tumors. The cystic lesions were diffusively located on the surface of the greater omentum, peritoneum, mesentery of the small intestine, and organs including the liver and spleen. No invasion of the intestinal wall or parenchyma of the liver and spleen occurred. Therefore, the final diagnosis was peritoneal cavernous hemangiomatosis.

Although a huge cystic lesion was located in the abdominal cavity, the platelet count and coagulation function tests were within the reference ranges. Thus, Kasabach–Merritt syndrome was excluded[10]. We also excluded Maffucci's syndrome because no deformity or activity limitation of the extremities was found[11]. Finally, we excluded Klippel–Trenaunay syndrome because the patient had no nevus vascularis, lower extremity deformity, or superficial varicosities[12].

No clear treatments for peritoneal hemangiomatosis have been established. We treated the patient with hyperthermic perfusion because of the malignant clinical manifestation of the tumor. However, the patient developed palpitation after the hyperthermic perfusion. Consequently, we stopped the hyperthermic perfusion treatment. Propranolol can be used to treat gastrointestinal hemangiomatosis[13] and hepatic hemangiomatosis[14]. We believe that propranolol may be an effective agent to treat the disease such as that described in the present case.

**CONCLUSION**

The possibility of peritoneal cavernous hemangiomatosis should be considered when multiple cystic masses are found in the abdominal cavity by preoperative examination.

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**Figure 1 Clinical, pathological and immunohistochemical results.** A: The white arrows indicate the abdominal masses; B: The inner cystic wall resected during the operation; C: CD31 staining was positive for the endothelial cells. Magnification × 100.