Case Report

Malignant Hemangiopericytoma of the Sigmoid Mesocolon Presenting as Acute Abdomen: Case Report and Review of the Literature

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Key Words
Hemangiopericytoma;
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Hemangiopericytoma is a rare solitary vascular tumor originating in the pericytes surrounding capillaries. It arises extremely rare in the abdomen, especially the mesocolon. There were only three cases reported in the literatures. Experience of treatment with this tumor is limited due to their rare occurrence and various malignant potential. We report the case of a 75-year-old female who the acute abdomen was caused by the malignant hemangiopericytoma of the sigmoid mesentery. She underwent surgical excision and adjuvant radiotherapy. Postoperative follow-up of 3-year duration revealed no recurrence.
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Hemangiopericytoma is an unusual solitary vascular tumor arising from the pericytes surrounding capillaries. Pericytes are rudimentary cells that have contractile properties and regulate blood flow through capillaries.[1] Hemangiopericytoma appears most commonly as a painless slow-growing mass arising from the soft tissue of lower extremities.[2] It arises extremely rare in the abdomen, with only a few cases reported to arise in greater omentum, colon, and mesentery.[3-7] The prognosis is uncertain due to its limited occurrence and various malignant potential. However, it had been recognized the ability for local recurrence and distant metastasis.[2] Here we report a patient with hemangiopericytoma originating in the sigmoid mesocolon managed by surgical excision and adjuvant radiotherapy.

Case Report

A 75-year-old female presented to our gynecologic section with progressive lower abdominal pain with a 3 days' duration. She reported no change in bowel habit, bloody stool, or weight loss. She had no systemic diseases. Two years ago, she was discovered a pelvic mass after the investigation for her occasional abdominal pain. At that time, computed tomography (CT) was performed and showed a low density soft tissue mass ($5.2 \times 4.0 \times 3.3$ cm^3) over the posterior aspect of lower pelvic region without contrast enhancement, which ovarian tumor was considered (Fig. 1). But she refused surgery and loss follow-up. On this admission, her vital signs were stable with a body temperature of 37.0 °C. Physical examination revealed

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local tenderness over left lower abdomen with muscle guarding. Laboratory data showed only normocytic anemia (hemoglobin value of 11.6 g/dL) and plain films were unremarkable. Abdominal ultrasound showed a large heterogeneous pelvic mass just near the uterus. She underwent a non-elective laparotomy due to the development of rebounding tenderness of the abdomen.

At operation, a reddish brown solid tumor about $12 \times 12 \times 10$ cm^3 in size was located at sigmoid mesocolon. The tumor had focal areas of hemorrhage and necrosis. It was very close to the colon wall and did not involve the loops of the bowel and other pelvic organs. There was no ascites or metastatic lesions. The coloproctologist was consulted during the operation and the Hartmann's procedure was performed under an unprepared colon. Microscopically, the sections showed pictures of anasplastic oval to spindle tumor cells. Some of the vessels were branching and exhibit a characteristic "staghorn" configuration. Focal area displayed fibrosis and hyalinization. The anaplastic cells had prominent mitotic activity, and marked hypercellularity. The immunohistochemical stains of the tumor cells showed positive staining for CD34 and Vimentin, and negative for actin, S-100, cytokeratin, desmin, CD10, and c-kit (Fig. 2A-E). These findings confirmed the diagnosis of a hemangiopericytoma arising from the sigmoid mesentery. The postoperative course was uneventful. Due to the aggressive malignant behavior in histopathologic findings, she was suggested to receive subsequent radiotherapy of the pelvis. The patient underwent the reversal operation for the restoration of intestinal continuity 4 months later. There was no evidence of recurrence or metastasis by imaging studies for 3 years following the initial operation.

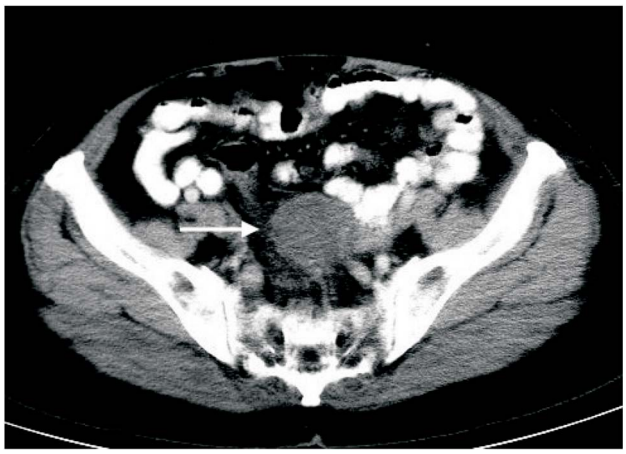

Fig. 1. Computed tomography (two years previously) showed a low density soft tissue mass ($5.2 \times 4.0 \times 3.3$ cm^3) over the lower pelvic region without contrast enhancement (arrow).

Discussion

Hemangiopericytomas are rare solitary vascular tumors representing less than 1% of vascular neoplasms.[2] They usually affect the third to sixth decade of adults with an equal distribution in male and female.[2] Lower extremities, retroperitoneum, pelvic fossa, head, neck, and chest are common sites of its occurrence.[2] In the review of English literature, only three cases of hemangio-

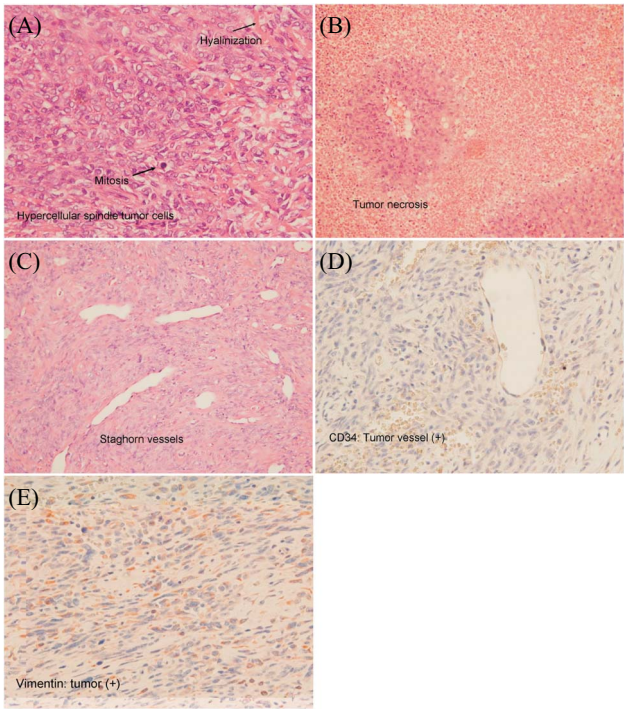

Fig. 2. (A) Anaplastic oval to short spindle tumor cells with high cellularity, abnormal mitoses, and focal area of hyalinization (hematoxylin and eosin, × 400). (B) Focal area display necrosis (hematoxylin and eosin, × 100). (C) Some of the vessels are branching and exhibit a characteristic "staghorn" configuration (hematoxylin and eosin, × 200). (D & E) Immunohistochemical stain for tumor cells showed positive reaction with CD34 and vimentin (× 400).

pericytoma arising from sigmoid mesentery had been reported.[5-7] Those three patients were all asymptomatic and were found the tumor incidentally for unrelated conditions. They received only surgical excision and the histopathologic results were all without evidence of mitotic figures. There was no recurrence occurred during the period of follow-up in these three patients.

Imaging findings are non-specific and there are no serum tumor markers indicating the presence of hemangiopericytoma. These cause an accurate preoperative diagnosis difficulty. Frozen section biopsies can not make definite diagnosis because the need for further immunohistochemical stains.

The diagnosis of hemangiopericytoma is mainly based on the results of histological features and immunohistochemical stains. Macroscopically, hemangiopericytoma is a well circumscribed or thinly encapsulated tumor. It is often fragile covered by a variable number of enlarged vessels that easily causing hemorrhage and making surgery difficult.[5,7] Microscopically, it is characterized by anaplastic oval to short spindle tumor cells around thin walled, endothelial lined vascular channels. Focal area display fibrosis and hyalinization. Some of the vessels exhibit a characteristic "staghorn" configuration.[7] These anaplastic cells may have mitotic activity, necrosis, hemorrhage and marked hypercellularity. Immunohistochemical (IHC) stains are useful for an accurate diagnosis. Hemangiopericytomas showed positive staining for CD34 and Vimentin, and negative for actin, S-100, cytokeratin, desmin, CD10, and c-kit.[5,7] The results of IHC stains of our presenting case showed positive to CD34 and vimentin, which indicates its vascular and mesenchymal origin, and negative to other markers including c-kit (CD117), cytokeratin, desmin, S-100 and CD10, which can exclude muscular or epithelial origin of the tumor such as gastrointestinal stromal tumor (GIST), melanoma or carcinoma. The main differential diagnosis of hemangiopericytoma is hemangioendothelioma. However, the unique "staghorn" configuration of some of the vessels in histological features is the only characteristic of hemangiopericytoma, and can exclude the differential diagnosis of hemangioendothelioma.

Hemangiopericytomas are classified as benign or malignant. It is important to evaluate the malignant potential of the tumor which is related to the recurrence or metastasis after resection. Tumor size more than 5 cm, a high mitotic activity with more than four mitoses per ten high power fields, hypercellularity, and hemorrhage and necrosis within the tumor are all possible prognostic indices to predict a malignant hemangiopericytoma.[8] The liver, lung, and peritoneum are common sites of recurrence.[3] Surgical excision is the mainstay of treatment for a disease-free survival.[9] Radiotherapy had been reported to be beneficial for those aggressive tumors or for those unresectable hemangiopericytoma as well as recurrence.[10] The effect of chemotherapy for this tumor has not been established.[3]

This case represents a case of hemangiopericytoma arising from sigmoid mesocolon with aggressive malignant behavior in histopathologic features as well as a clinical presentation of acute abdomen which had never been described in the literature. She underwent surgical excision followed by adjuvant radiotherapy. Postoperative follow-up of 3-year duration revealed no recurrence. Due to an uncertain prognosis, a long term follow-up is necessary because disease relapse may occur after long periods of time.[2]

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病例報告

乙狀結腸繫膜之惡性血管外皮細胞瘤表現出急性腹症：病例報告與文獻回顧

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惡性血管外皮細胞瘤是一個少見而單獨的血管腫瘤，起源於微細血管旁的外膜細胞。它發生在腹部是非常罕見的，尤其是在結腸繫膜，只有三例個案在文獻上被報告過。由於少見及具有不同的惡性度，目前為止對於這個腫瘤的治療經驗仍十分有限。我們報告一位七十五歲的女性，由於乙狀結腸腸繫膜上的惡性血管外皮細胞瘤而表現急性腹症。她接受了手術切除及輔助性放射治療。術後追蹤三年期間無復發情形。

關鍵詞 血管外皮細胞瘤、腸繫膜、乙狀結腸。