Clinical Case Report

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Peritoneal epithelioid angiosarcoma

A case report

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Abstract

Introduction: Peritoneal angiosarcoma is an extremely rare sarcoma (0.01287% incidence per 100,000) with an aggressive course and a poor prognosis. In this case, the manifestation of peritoneal angiosarcoma was ascites, which caused difficulty in early diagnosis and the diagnosis of peritoneal angiosarcoma, was made only after the surgery.

Patient concerns: A 61-year-old man working in Mainland China presented with a 1-month history of abdominal distension. A contrast-enhanced abdominal computed tomography (CT) scan revealed peritoneal carcinomatosis with massive ascites. However, his tuberculosis (TB) polymerase chain reaction was negative. The ascites cell block and cytology also revealed negative for malignant cells. The patient underwent intra-abdominal tumor excision. After the operation, the patient's blood pressure (BP) dropped. Due to the state of shock, he was transferred to an intensive care unit (ICU).

Diagnoses: According to the pathology report, the neoplastic cells were positive for cytokeratin, cluster of differentiation 31 (CD31), cluster of differentiation 34 (CD34), and negative for cytokeratin 7 (CK7), cytokeratin 7 (CK20). Therefore, the diagnosis of epithelioid angiosarcoma was made.

Interventions: The patient took 400 mg of Pazopanib once a day.

Outcomes: Even though vasopresser was used, the patient's BP was still low. Finally, he expired.

Lessons: Initialy, the patient presented with abdominal distension and large amount of ascites in the beginning. TB peritonitis was highly suspected after the abdominal CT scan. Therefore, surgical procedures would be essential in the identification of proper diagnosis. In the future, the diagnosis of peritoneal epithelioid angiosarcoma should also be taken into consideration for patients with abnormal ascites besides the common diagnoses of TB, liver cirrhosis, and infection.

Abbreviations: BP = blood pressure, CD31 = cluster of differentiation 31, CD34 = cluster of differentiation 34, CK20 = Cytokeratin 7, CK7 = cytokeratin 7, CT = computed tomography, EGD = esophagogastroduodenoscopy, TB = tuberculosis.

Keywords: angiosarcoma, ascites, epithelioid angiosarcoma, peritoneum, sarcoma

1. Introduction

More than 50 histological types of soft tissue sarcomas have been identified by the World Health Organization classification. Angiosarcomas which belong to a subtype of soft tissue sarcomas are rare and aggressive vascular malignancy of endothelial origin.[1,2] They most frequently occurs for the skin of the head and neck region or the extremeties.[2,3] Regardless of tumor origin, the prognosis remains poor with an overall 1-year survival rate of 50% and 5-year survival rate of just 35%.[2,4] Peritoneal angiosarcoma is an extremely rare sarcoma (0.01287% incidence per 100,000) with an aggressive course and a poor prognosis compared with the more typical origins on the head or extremities in the literature.[5] We report the case of epithelioid angiosarcoma of the peritoneum.

2. Case presentation

A 61-year-old man working in Mainland China presented with a 1-month history of abdominal distention after taking a meal. The persistent symptom caused his abdominal cavity to look obviously bloated or enlarged. However, he had no history of diabetes, hypertension, hepatitis, or tuberculosis (TB). Besides, he not only had no exposure to drugs, toxin, or radiation but also did not suffer from weight loss. He further denied orthopnea, lower-limb edema, decreased urinary output, and jaundice. Systemic examination was unremarkable except for positive shifting dullness and slight tenderness. Original laboratory tests revealed glutamic-pyruvic transaminase 14 IU/L (10–40), glutamic-oxalocetic transaminase 21 IU/L (10–42), total bilirubin 0.54 mg/dL (0.2–1.0), and direct bilirubin 0.1 mg/dL (0–0.2). Tumor markers including alpha-fetoprotein 2.19 (<9.0 ng/mL), carcinoembryonic antigen 1.76 (0–5 ng/mL), and carbohydrate antigen 199 35.52 (<37 U/mL) were all within normal limits. Serum surface antigen for hepatitis B and anti-hepatitis C virus were negative. The initial renal function test was normal. The albumin value was 3.47 g/dL (3.5–5.0).

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none.

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The patient's chest x-ray showed nonspecific findings. Subsequently, both esophagogastroduodenoscopy (EGD) and colonoscopy were done. The EGD showed reflux esophgitis and chronic gastritis. The colonoscopy revealed internal hemorrhoid.

An abdominal ultrasonography revealed a large volume of ascites, so paracentesis was performed. The analysis showed cell counting: 890 cells/mm^3, PMN 3%, mono 97%, and albumin 2.72 g/dL.

A contrast-enhanced abdominal computed tomography (CT) scan revealed peritoneal carcinomatosis with massive ascites (Fig. 1A). There is doubt whether this patient would be diagnosed with TB or malignancy. However, his TB polymerase chain

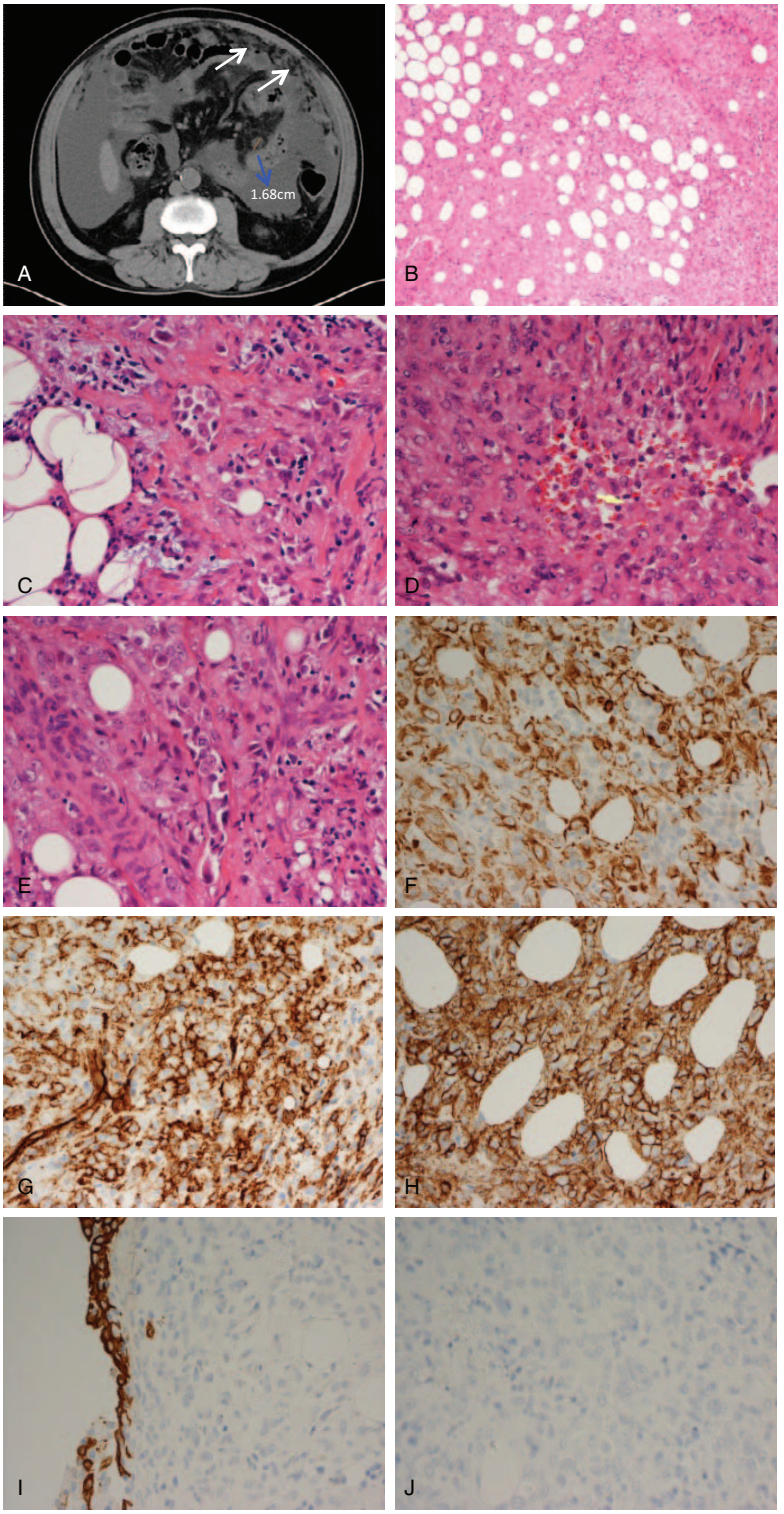

Figure 1. (A) Computed tomography scan of the abdomen demonstrated peritoneal carcinomatosis thickening (white arrows) and fluid accumulation in the abdomen. The largest tumor was about 1.68 cm (blue arrow) (B) 100 ×, (C) 400 ×, (D) 400 ×, (E) 400 ×, (F) CK(+), (G) CD31(+), (H) CD34(+), (I) CK7(−), (J) CK20(−).

reaction was negative. The ascites cell block and cytology also revealed negative for malignant cells.

On the basis of the previous diagnosis, the patient underwent intra-abdominal tumor excision. During the operation, carcinomatosis was found on the omentum, and massive ascites was drained up to 7600 cc. After the operation, the patient's blood pressure (BP) dropped, and his hemoglobin was 4.3 g/dL (13.4–17.2). Owing to the state of shock, he was transferred to an intensive care unit.

According to the pathology report, the tumor was composed of solid sheets of high-grade epithelioid neoplastic cells with abundant amphophilic to lightly eosinophilic cytoplasm, large vesicular nuclei, and prominent nucleoli. Focal complex anastomosing vascular channels and brisk mitoses were noted (Fig. 1B–E). The neoplastic cells were positive for cytokeratin (CK, Fig. 1F), cluster of differentiation 31 (CD31) (Fig. 1G), CD34 (Fig. 1H), and negative for CK7 and CK20 (Fig. 1I and J). In addition, on the basis of morphological features and immunohistochemical results of the tumor, the diagnosis of epithelioid angiosarcoma was made.

Although the patient's massive bloody ascites was drained out after the operation, the symptom of his abdominal distention did not ameliorate for the continual occurrence of ascites. For the removal of ascitic fluid, paracentasis was performed in ICU. After the paracentasis, he started taking 400 mg of Pazopanib once a day. Even though vasopresser was used, his BP was still low. Finally, the patient expired. This study was approved by the Ethics Committee of the Kaohsiung Medical University Hospital and the signed informed consent was obtained from the patient for publication of this report.

3. Discussion

Pathologic diagnosis of angiosarcoma often requires thorough staining, particularly in poorly differentiated tumors. The subtypical epithelioid angiosarcomas are generally positive for CD31 and variably positive for CD34.[6] In contrast, a carcinoma would be positive for CK, but negative for CD31 and CD34.[6] In our case, the neoplastic cells were positive for CK, CD31, and CD34.

The rarity of peritoneal angiosarcoma makes it worthy of attention. In this case, the patient presented with abdominal distention and large amount of ascites in the beginning. Besides, the EGD and colonoscopy showed no malignancy. Consequently, TB peritonitis was highly suspected after the abdominal CT scan. Although further ascitic fluid analysis and cytology were negative finding of TB, the sensitivity of an ascitic smear for diagnosis of TB peritonitis is only 2.93%.[7] In addition, the sensitivity of an ascitic fluid culture is 34.75%.[7] However, the sensitivity of laparoscopy is 93%.[7] Therefore, surgical procedures would be taken into account and be analyzed in the identification of proper diagnosis.

Evidence-based treatments of angiosarcomas are scarce.[8] According to Seo et al,[8] radical resection is considered for radiation-induced angiosarcomas, and systemic chemotherapy is for inoperable, locally advanced or metastatic angiosarcomas. However, in our case, the patient was in the state of shock after the operation, so the surgical intervention and the chemotherapy were comparably unsuitable treatments. Therefore, we prescribed tyrosine kinase inhibitor such as Pazopanib for this patient by oncologist's suggestion.

The limitation of this study is that the peritoneal epithelioid angiosarcoma is quite rare, and the diagnostic method and the treatment are seldom recorded in the literature. In the future, the diagnosis of peritoneal epithelioid angiosarcoma should also be taken into consideration for patients with abnormal ascites besides the common diagnoses of TB, liver cirrhosis, and infection. According to this case, once primary tumors are not found after related examinations such as EGD and colonoscopy, surgical procedures should be performed as soon as possible. As long as the definite diagnosis is made, target therapy will be offered in advance.

Author contributions

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References

[1] Dei Tos AP, Wagner AJ, Modena P, et al. Epithelioid soft tissue tumors. Semin Oncol 2009;36:347–57.
[2] Young RJ, Brown NJ, Reed MW, et al. Angiosarcoma. Lancet Oncol 2010;11:983–91.
[3] Greenwald U, Newman E, Taneja S, et al. Malignant epithelioid angiosarcoma of the external iliac vein presenting as venous thrombosis. Ann Vasc Surg 2004;18:493–6.
[4] Pazona JF, Gupta R, Wysock J, et al. Angiosarcoma of bladder: long-term survival after multimodal therapy. Urology 2007;69:9–10. 575 e.
[5] Litvan J, Aghazarian M, Wiley E, et al. Primary peritoneal angiosarcoma: a case report. Anticancer Res 2014;34:5001–6.
[6] Hart J, Mandavilli S. Epithelioid angiosarcoma: a brief diagnostic review and differential diagnosis. Arch Pathol Lab Med 2011;135:268–72.
[7] Sanai FM, Bzeizi KI. Systematic review: tuberculous peritonitis—presenting features, diagnostic strategies and treatment. Aliment Pharmacol Ther 2005;22:685–700.
[8] Seo CJ, Lek SM, Tan GHC, et al. Radiation-associated peritoneal angiosarcoma. BMJ Case Rep 20172017.