

Case Report

Acute Abdomen Caused by Ruptured Angiomatoid Fibrous Histiocytoma

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SUMMARY

Angiomatoid fibrous histiocytoma (AFH) is a rarely seen soft tissue tumor often affects children or young adults. The tumor possesses intermediate behavior and usually arise in superficial soft tissue in trunk and extremities, visceral organs such as the ovary, vulva, brain, and lung. We present a 77-year-old old woman who suffered from a sudden onset of abdominal pain with hypovolemic shock. Vital signs showed tachycardiac hypotension (blood pressure: 80/64 mmHg; heart rate: 106/minute). On physical examinations, patient presented with pale conjunctiva, diffuse abdominal tenderness and rigidity. Bedside sonography shows ascites with heterogeneous echogenicity and a cystic lesion with 2.88 cm in diameter in left upper quadrant (LUQ) abdominal area. Under the impression of hemoperitoneum caused by a ruptured vascular lesion, computed tomographic angiography (CTA) was performed and revealed one 2.5 cm cystic lesion with extravasation of contrast medium raising concern of ruptured space-occupying lesion with hemoperitoneum. Laparoscopic hemostasis by the general surgeon was performed. After resection the cystic lesion, the patient recovered smoothly and discharged 10 days after surgery. The definite pathologic diagnosis is AFH. This is the first case describing intra-abdominal AFH with acute abdomen and hemorrhagic shock.

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1. Introduction

Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue tumor often affects children or young adults.¹ The tumor is intermediate behavior and usually arise in superficial soft tissue in trunk and extremities, visceral organs such as the ovary, vulva, brain, and lung are also recognized locations for this tumor.^{2,3} We present an old woman presented as sudden onset abdomen pain with peritoneum sign and hypovolemic shock. Sonography and contrast-enhanced computed tomography angiography (CTA) present a leaking cystic lesion abutting stomach great curvature and hemoperitoneum. After resuscitation and successful surgical resection, the patient was recovery smoothly and discharged. Definite pathologic diagnosis is AFH. This is the first case describing AFH omentum presenting with hemorrhagic shock. We also report the clinical challenges before a definite diagnosis was made.

2. Case report

A 77 years old female has a history of well-controlled type 2 diabetes and hypertension. She suffered from sudden onset epigastric pain and near fainting 6 hours prior visiting our emergency department. Vital signs showed hypotension with narrow pulse pressure and tachycardia (blood pressure: 80/64 mmHg; heart rate: 106/minute). On physical examination, the patient presented with

pale conjunctiva, abdomen rigidity, diffuse tenderness and rebounding pain. Immediate intravascular (IV) access and volume supplement was administrated. Bedside sonography shows ascites with mixed internal echogenicity, a cystic lesion with 2.88 cm in diameter located in the left upper quadrant abdomen area with posterior acoustic enhancement (Figure 1a). Under the impression of hemoperitoneum related to vascular lesion rupture, we performed computed tomographic angiography (CTA) after adequate IV fluid supplement and blood transfusion. CTA showed one 2.5 cm cystic lesion with extravasation of contrast medium and high attenuated fluid collection abutting greater curvature of stomach; moderate amount high attenuation ascites raising the concern of hemoperitoneum (Figure 1b). After discussion with a radiologist and general surgeon, angiography embolization was applied first but failed to locate the leaking vascular lesion. Under the impression of undetermined etiology cystic lesion induced internal bleeding, laparoscopy for diagnosis and stop bleeding was performed. Intraoperative findings include a massive amount of bloody ascites about 2000 cc in abdominal cavity and a 3 cm cystic tumor between greater curvature and splenic flexure of colon (Figure 1c). After resection the cystic lesion, patient recovered smoothly and discharged 10 days after surgery.

Pathology specimen showed a cystic lesion measured 3.2 cm in dimension with blood clots and dark-red component. Microscopically, hematoxylin and eosin (H-E) stained sections show soft tissue with a relatively well circumscribed cystic lesion. The lesion is composed of some areas of the spindle or histiocytoid cells with a syncytial growth and other areas show pseudoangiomatous spaces

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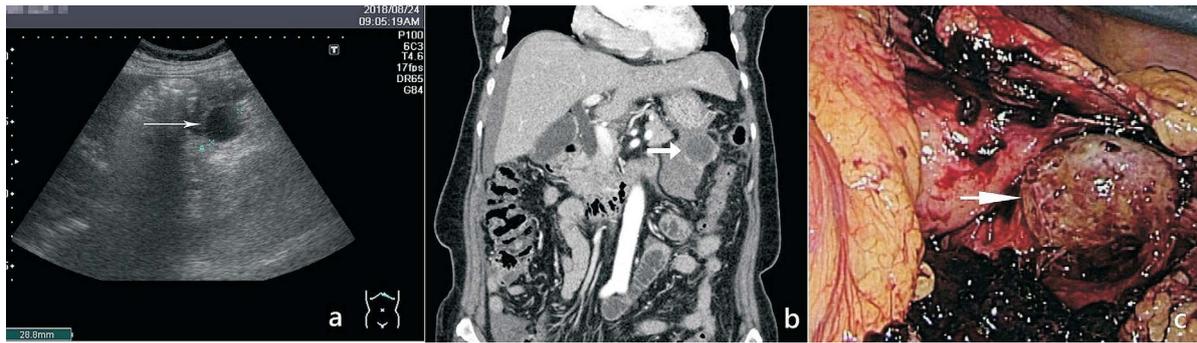


Figure 1. (a) A cystic lesion with 2.88 cm in diameter located in the left upper quadrant abdomen area with posterior acoustic enhancement. (b) CTA showed one 2.5 cm cystic lesion (arrow) with extravasation of contrast medium abutting greater curvature of the stomach. (c) A 3 cm cystic tumor (arrow) between greater curvature and splenic flexure of colon.

filled with blood. AFH in low-magnification microphotography originated from omentum is showed in H-E stain at 10x (Figure 2D). A thick fibrous pseudo-capsule with hemosiderin deposition is also observed. Pericapsular cuffing of lymphoplasmacytic cells is occasionally seen (Figure 2). The spindle or histiocytoid cells are CD99 (+), CD68 (focal+), CD31 (focal+), smooth muscle actin (SMA) (focal+), CD34 (-), ERG (-), HHV-8 (-), epithelial membrane antigen (EMA) (-), CK (AE1/AE3) (-), and desmin (-) (Figure 3). The morphology and immunoprofile are most consistent with that of an angiomatoid fibrous histiocytoma (AFH).

3. Discussion

Accurate diagnosis in elderly patients with gastrointestinal (GI) symptom is challenging. In a retrospective study in Japan, primary care physicians visiting elderly patients with GI symptoms at home only yield 33.3% diagnosis accuracy.⁴ AFH is a rare soft tissue tumor usually found in children and young adult arising in dermal and subcutaneous tissue. It accounts for approximately 0.3% of soft-tissue neoplasms.⁵ The first diagnosis of AFH was made in 1979 by Enzinger as a subtype of malignant fibrous histiocytoma.¹ Subsequent larger studies demonstrated a markedly lower rate of malignancy than observed in the original series of 41 patients, and the lesion thereafter was renamed AFH.^{6,7} This rare tumor shares histologic and radiographic features with other fibrous lesions, thus leading to its diagnostic difficulty. Majority of the cases occur in the

extremities with clinical symptoms present as a recurrent bleeding lesion in an identical location. Recently, AFH has been reported in unusual sites including the lung, mediastinum, vulva, retroperitoneum, ovary, pulmonary artery, kidney, omentum, and intracranial.⁸⁻¹⁶

Although AFH was thought to be relative noninvasive soft tissue tumor, the recurrent rate, and outcome vary based on previous case reports. Recurrent rate varies from 0 to 63%, metastasis rate from 0 to 29%, and mortality rate from 0 to 14.3%.^{1,6,7,9,17-21} Several observation findings suggest poor prognosis by the previous reports. Costa et al. state an irregular tumor border involving head or neck location were associated with higher local recurrence, and the depth of the tumor was correlated with subsequent local and distant metastasis.⁶ Saito K. et al. have found some specific finding in their two mortality cases: The first case suffered from a relative painful mass initially, and microscopically exhibited an unusual small round cell morphology with marked mitotic activity. The second case manifested clinically as a large mass in a deeper intramuscular tissue, and its recurrence tumor demonstrated focally increased nuclear atypia and pleomorphism compared to the classic morphology of primary lesion.²¹

According to the past reports, the patient is clinically life-threatening from primary tumor only in intracranial AFH bleeding.^{11,14,15,22} This is the first case presenting AFH bleeding with hemoperitoneum and hypovolemic shock. The initial resuscitation based on bedside sonography is crucial to stabilize patient. Under the impression of hemoperitoneum caused by unknown etiology, emergent CTA and then embolization for definite diagnosis was arranged. Finally, the

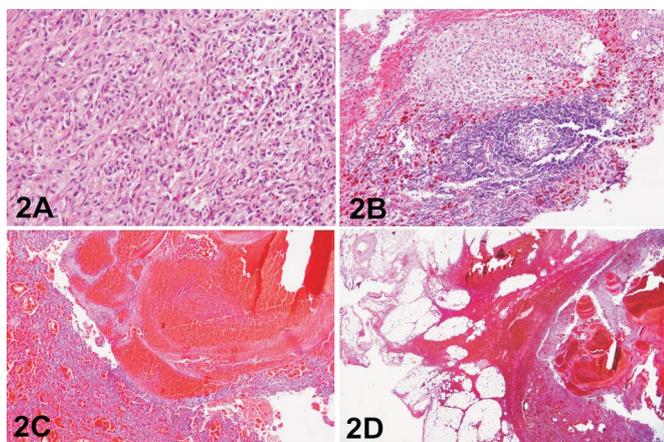


Figure 2. Soft tissue with a relatively well circumscribed cystic lesion, H-E stain at 200x (A). Pericapsular cuffing of lymphoplasmacytic cells, H-E stain at 100x (B). Lesion is composed in some areas of spindle or histiocytoid cells with a syncytial growth and other areas show pseudoangiomatous spaces filled with blood, H-E stain at 40x (C). Relationship of omentum and AFH in low-magnification microphotography showed in H-E stain at 10x (D).

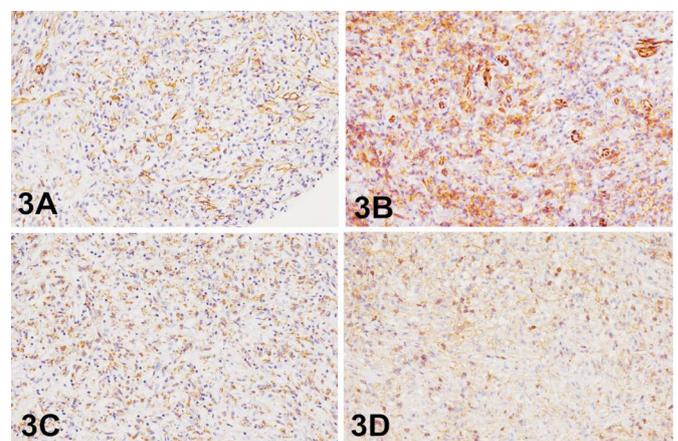


Figure 3. The spindle or histiocytoid cells in SMA immunostain at 200x (A). CD31 immunostain at 200x (B). CD68 immunostain at 200x (C). CD99 immunostain at 200x (D).

surgeon performed laparoscopic resection after the the embolization was failure. The patient was fairly recovered after surgery and discharged from the ward. Follow up in out-patient-department 2 months later showed no sign of abdomen pain and internal bleeding.

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Declarations of interest

None.

Author contributions

All authors had access to the data. Yang HW wrote the draft and gathered data. Su YJ revised and corresponded. Chang WC provided pathology film and discussion.

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