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Case Report

Unusual Presentation of Peritoneal Angiomatoid Fibrous Histiocytoma in an Elderly Female

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SUMMARY

Angiomatoid fibrous histiocytoma (AFH) is a rare soft tissue tumor with low-grade malignant potency that has typically occurred in the extremities of young adults over the last 30 decades. A 77-year-old female presented to the emergency department with a sudden onset of severe abdominal pain. Contrast-enhanced computed tomography demonstrated a 2.5 cm cystic tumor in the upper abdomen that appeared ruptured with hemorrhage. The tumor that arose from the peritoneum was successfully resected and proven to be a ruptured AFH. The patient remained recurrence-free at the 1-year follow-up.

1. Introduction

Angiomatoid fibrous histiocytoma (AFH) is a rare low-grade soft tissue neoplasm that accounts for 0.3% of all soft tissue tumors. Most cases of AFH present in the first three decades of life.[1,2] It predominantly arises in the superficial areas of the extremities in children or young adults. It mainly occurs in the deep dermis and subcutaneous tissue and usually shows slow growth. AFH seldom causes symptoms. A few patients experience systematic symptoms such as anemia, easy bleeding, pyrexia, and malaise, suggesting tumor cytokine production.[3] Diagnosis is based on pathological review and immunochemistry. Wide local excision is suggested for patients with symptoms associated with AFH.[4,5] We reported an intra-abdominal AFH arose from an unusual location in the peritoneum with bleeding in an elderly female.

2. Case report

A 77-year-old female with a history of hypertension complained of sudden onset abdominal pain and fullness. She denied having fever, constipation, weight loss, hematemesis, melena, or hematochezia. Upon examination in the emergency department, she seemed uncomfortable, and the vital signs indicated shock. The systolic blood pressure was 60 mmHg. Tachycardia, which was 110 bpm, was noticed at the same time. The abdomen was distended with active bowel sounds. On palpation, diffuse abdominal tenderness with guarding and rebound were observed. Lymphadenopathy was not palpable. A digital rectal examination yielded brown stool that tested negative for blood. The white blood cell count was 11,600/uL and hemoglobin was 10.6 g/dL. The results of the biochemical panel and coagulation tests were within normal limits. The chest and plain abdominal radiographs were normal. Endogastroduodenoscopy revealed a shallow gastric ulcer without bleeding. Contrast-enhanced computed tomography (CT) showed the possibility of hemoperitoneum and a 2.5 cm cystic lesion with extravasation of contrast medium and highly attenuated fluid collection situated between the greater curvature of the stomach, splenic flexure of colon and left side kidney; it was supplied by two vessels from peritoneum (Figure 1A-C). Abdominal ultrasonography also revealed bloody ascites diagnosed by paracentesis. The soft tissue tumor, arising from the peritoneum, situated within the omental bursa, positioned below the tail of the pancreas and interposed between the greater curvature of the stomach and the splenic flexure of the colon, was excised (Figure 1D). During the operation, a massive amount of bloody ascites, more than 2000 ml, was accumulated in the abdominal cavity. Histopathological evaluation of the resected specimen revealed spindle-to-epithelioid cells with a syncytial growth pattern, with areas showing pseudoangiomatous spaces filled with blood. A thick fibrous pseudocapsule with hemosiderin deposition was also observed (Figure 2). On immunohistochemical examination, CD68, CD31 and smooth muscle actin (SMA) were positive, and desmin was negative (Figure 3). It was confirmed to be a ruptured AFH with hemorrhage. The patient remained recurrence-free at the 1-year follow-up.

3. Discussion

The most commonly diagnosed cancers worldwide have been breast, lung, and prostate cancers.[6] Old age plays a vital role in their incidence.[7–9] However, AFH is an unusual soft tissue neoplasm that occurs mostly in children or young adults. To the best of our knowledge, no elderly patient has been diagnosed with AFH in the peritoneum until now.

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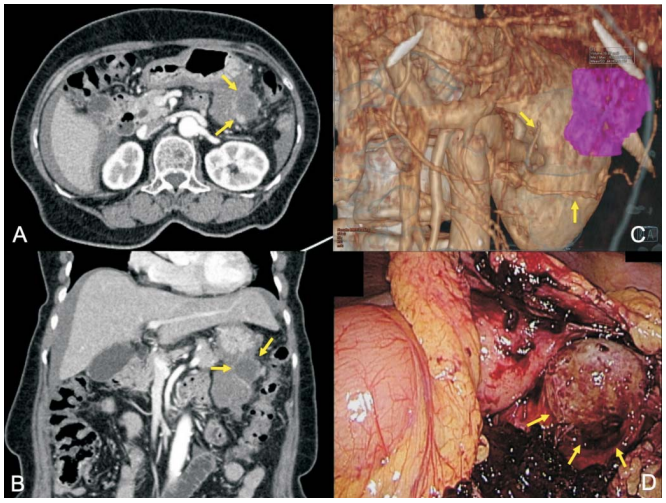

Figure 1. A: Post-enhanced computed tomography shows a 2.5 cm cystic lesion with extravasation of contrast medium and high attenuated fluid collection situated between stomach and left side kidney (arrow). B: In the sagittal view of post-enhanced computed tomography, the cystic lesion located between the great curvature of the stomach and splenic flexure of the colon (arrow). C: In the three-dimensional reconstructed image, the cystic lesion (pink area) was supplied by two surrounding vessels (arrow). D: Intraoperative photograph of a 2.5 cm cystic tumor was situated within the omental bursa, positioned below the tail of the pancreas and interposed between the greater curvature of the stomach and the splenic flexure of the colon. It also exhibited adherence to surrounding tissues.

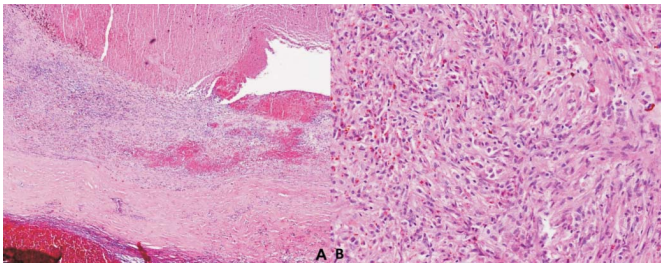

Figure 2. Photomicrograph of the histopathologic evaluation showed A: Spindle or histiocytoid cells with a syncytial growth and other areas show pseudoangiomatous spaces filled with blood. Pseudocapsule with hemosiderin deposition is observed. B: Spindle to epithelioid cells in syncytial growth, admixed with hemosiderin deposition (HE stain, magnification ×40 in [A], ×200 in [B]).

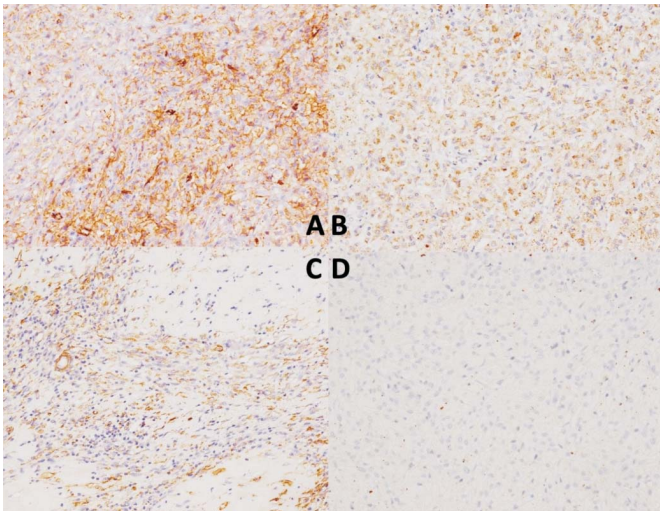

Figure 3. Photomicrograph of immunohistochemical evaluation showing the spindle or histiocytoid cells. A: Positive for CD31, B and C: Focal positive for CD68 and smooth muscle actin (SMA), D: It was negative for desmin (magnification × 200).

Differential diagnosis of intra abdominal tumors, including peritoneal tumors, is a complex process that requires a systematic approach combining patient history, physical examination, laboratory tests, and imaging studies. Biopsy and histopathology are often necessary for a definitive diagnosis.[10] The differential diagnosis for peritoneal tumors is diverse, encompassing a wide spectrum of conditions, from benign granulomas to malignant neoplasms.[10] These diagnoses can be categorized into two distinct groups based on their origin: primary and secondary. Primary peritoneal tumors are uncommon tumors originating from transformed cells in the peritoneal and subperitoneal layers. The subperitoneal space comprises a delicate layer of connective tissue housing adipose tissue, blood vessels, lymphatic vessels, and nerves. This region is known for giving rise to various primary mesenchymal neoplasms, such as cystic lymphangiomas, mesenteric lipomas, gastrointestinal stromal tumors, peritoneal liposarcomas, and the aforementioned angiomatoid fibrous histiocytoma.[11] These mesenchymal soft tissue tumors can initially be distinguished by characteristics including their location, distribution pattern, and texture when examined via computed tomography. Nevertheless, confirmation through histopathological examination stands as the definitive method for diagnosis.[11] Secondary peritoneal tumors, known as peritoneal carcinomatosis, are more prevalent and result from the spread of metastatic cancers, typically from abdominal neoplasms like ovarian and gastrointestinal cancers. Occasionally, non-abdominal cancers like breast, lung, or melanoma can also metastasize to the peritoneum.

AFH is a hypervascular tumor supported by multiple surrounding vessels. Hence, varying degrees of tumor hemorrhage can develop spontaneously or as a result of chemotherapy. Approximately 5% of AFHs will develop apparent intra-tumor hemorrhage spontaneously, especially when AFH arises from the peritoneum or omentum. A previous report described intraperitoneal AFH accompanied by apparent intra-abdominal hemorrhage, making diagnosis difficult. Chen, et al. reported an AFH case mistaken for ruptured hepatocellular carcinoma in 2007.[12] However, angiography confirmed that the mass was supplied by the arteries of the omentum instead of the hepatic artery. The above evidence shows that the tumor originated from the peritoneum instead of the liver.

The diagnosis of AFH is based on histopathology and immunohistology.[1,13] Under microscopy, the characteristic appearance includes the distribution of ovoid to spindle cells with bland, vesicular nuclei, and lymphoplasmacytic infiltrate with a fibrous pseudocapsule.[14] Immunoreactivity for smooth-muscle actin and design was approximately 40–50%. CD99, CD68 and CD31 have been variably reported, ranging from approximately 40% to 100% of lesions.[16,17] Recently, cytogenetic analysis has been added to the diagnosis of AFH with *EWSR1-CREB1*, *EWSR1-ATF1*, and *FUS-ATF1* fusion gene presentations.[1]

A few patients with AFH also present with paraneoplastic symptoms such as platelet dysfunction, easy bleeding, bruising, and anemia due to systemic inflammatory effects. Cytokine analysis of two patients with inflammatory AFH demonstrated elevated interleukin-6 (IL-6) and tumor necrosis factor levels in one or both patients.[17] However, the above symptoms and IL-6 levels improved after complete tumor excision, indicating that IL-6 plays a vital role in paraneoplastic symptoms. AFH also increases local inflammatory cytokines and can reduce hemostasis and coagulation.[18] The transcriptional regulation of IL-6 includes nuclear factor-kB, cAMP response element-binding protein, and activator protein 1. Hence, the *EWSR1-CREB1* fusion gene leads to constant activation of *CREB1*, re-

sulting in continuous IL-6 production.

The cumulative findings of a meta-analysis of multiple studies demonstrated that the majority of patients (73.2%) were disease-free after local excision, and a minority (23.2%) developed recurrent disease.[4] Less than 5% had been reported to experience metastasis, predominantly to regional lymph nodes and exceptionally to the lungs, liver, or brain.[19] Huijuan, et al. also reported that only two patients (2/21 9.5%) had tumor recurrence at 3 months and 6 months after tumor resection. The other patients survived without recurrence or metastasis after a median duration of 48 months.[20]

In conclusion, AFH typically occurs in the deep dermis and subcutaneous tissues of the extremities. They rarely occur in the peritoneum. It is challenging to distinguish AFH from other lesions arising from the peritoneum by CT or magnetic resonance imaging (MRI). Pathological reviews play a crucial role in the diagnosis. A small number of AFH cases recur locally, and rare cases are known to metastasize. Here, we present an unusual case of peritoneal AFH in an elderly female. To our knowledge, this is the first case of peritoneal AFH occurring in an elderly female in her late seventies. It showed that AFH is an important differential diagnosis if the patient has hemoperitoneum.

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Declaration of interest

The authors declare that they have no conflict of interest.

Ethical approval

This case report was approved by the Institutional Review Board of the MacKay Memorial Hospital (IRB number 23MMHIS197e).

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