Metastatic splenic angiosarcoma presenting with anemia and bone marrow fibrosis mimicking primary myelofibrosis: A case report and literature review

MEIQING WU, ZHONGQING LI, LIN LUO, WEIHUA ZHAO and JUN LUO

Department of Hematology, The First Affiliated Hospital of Guangxi Medical University, Nanning, Guangxi 530021, P.R. China

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Abstract. Angiosarcomas, originating from endothelial cells, are infrequent soft tissue sarcomas characterized by a high propensity for metastasis and an unfavorable prognosis. Splenic angiosarcoma, an exceedingly rare and aggressive neoplasm, exhibits variable clinical manifestations. The present case report describes a patient initially exhibiting anemia and bone marrow fibrosis, mimicking primary myelofibrosis, ultimately diagnosed with splenic angiosarcoma. The findings of the present case report underscore the importance of considering splenectomy for histopathological confirmation. Employing a panel of vascular differentiation markers is invaluable for establishing the diagnosis of angiosarcoma.

Introduction

Angiosarcomas demonstrate diverse clinical presentations and can be frequently misinterpreted as benign vascular tumors or other non-vascular malignancies (1). Splenic angiosarcomas are exceptionally rare tumors with a poor prognosis (2). Anemia and bone marrow fibrosis are typically associated with advanced stages of myeloproliferative neoplasms and other secondary causes (3). Additionally, splenomegaly is a common hallmark of hematological disorders. The present study describes the case of a patient, in whom the initial symptoms of anemia and bone marrow fibrosis, suggestive of primary myelofibrosis, culminated in the ultimate diagnosis of splenic angiosarcoma following a splenectomy.

Correspondence to: Professor Jun Luo, Department of Hematology, The First Affiliated Hospital of Guangxi Medical University, 6 Shuangyong Road, Nanning, Guangxi 530021, P.R. China
E-mail: luojungz@medmail.com.cn

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Case report

Clinical presentation. In September 2021, a 35-year-old woman was admitted to the First Affiliated Hospital of Guangxi Medical University with progressively worsening fatigue and bone pain. A complete blood count upon admission revealed anemia, with a white blood cell count of $6.48x10^9$/l, hemoglobin at 69 g/l and platelet count of $156x10^9$/l. Serum chemistry tests indicated elevated erythropoietin levels [156 mIU/ml; normal range, 4.3-29 mIU/ml, chemiluminescent immunoassay (Siemens Healthcare Diagnostics Products Limited)], elevated lactate dehydrogenase [356 U/l; normal range, 109-245 U/l, lactate substrate method (Zhongsheng Beikong Biotechnology Co., Ltd.)] and increased ferritin levels [1,524.67 ng/ml; normal range, 4.06-204 ng/ml, chemiluminescent immunoassay (Abbott Ireland Diagnostics Division)]. Total bilirubin levels were increased at 32.6 μmol/l (normal range, 3.4-20.5 μmol/l), as were direct bilirubin levels at 20.8 μmol/l (normal range, 0-6.8 μmol/l) [vanadate oxidation method (Zhongsheng Beikong Biotechnology Co., Ltd.)], and albumin levels were reduced at 26.7 g/l [normal range, 40-55 g/l, bromocresol green method (Siemens Healthcare Diagnostics Inc.)]. Coagulation profiles demonstrated normal prothrombin time and activated partial thromboplastin time [coagulation method (Instrumentation Laboratory Co.)], decreased fibrinogen [0.79 g/l; normal range, 2-5 g/l, Clauss method (Instrumentation Laboratory Co.)], and elevated levels of fibrinogen degradation products [251.26 μg/ml; normal range, 0-5 μg/ml, immunoturbidimetry (Biokit S.A.)] and D-Dimer [40,346 ng/ml; normal range, 0-450 ng/ml, immunoturbidimetry (Instrumentation Laboratory Co.)]. The peripheral blood and bone marrow smears were stained using the Wright stain (Tianjin Guangfu Fine chemical Research Institute) for 20 min at room temperature, washed under running water, dried up at room temperature, and observed under a microscope (Olympus BX43, Olympus Corporation). Peripheral blood smear examination revealed a variety of irregularly shaped erythrocytes, including target-shaped, elliptic, teardrop-shaped, helmet-shaped, and spherical red blood cells (Fig. 1A). Erythroblasts were frequently observed in peripheral blood, with a nucleated red cell to white cell ratio of 66 to 100. Additionally, immature granulocytes

were presented in peripheral blood (Fig. 1B). Bone marrow smears exhibited 28% granulocytes and 56% erythrocytes, which were mainly late juvenile erythrocytes. There was no obvious proliferation or abnormal morphology of megakaryocytes. Bone marrow biopsy demonstrated no blast cell proliferation and mild myelofibrosis. Flow cytometry [flow cytometer DxFLEX (Beckman Coulter, Inc.), data analysis by CytExpert, Gate setting: CD45-SSC Gates] of the bone marrow revealed no abnormal immunophenotype cells. Bone marrow aspiration was sent to a qualified third-party company (Kindstar Globalgene Technology, Inc.) for related gene testing and karyotype analysis. The *BCR-ABL1* fusion gene, *JAK2* V617F and *MPL* W515L/K gene mutation were detected using polymerase chain reaction. *CALR* exon 9 and *JAK2* exon12 were detected using sequencing. Fusion gene analysis of *BCR/ABL* and mutational analysis of *JAK2, CALR* and *MPL* genes yielded negative results. The karyotype of bone marrow was normal (data not shown). Normal levels of glucoencephalosidase, Lyso-GL-1 biomarkers [tandem mass spectrometry (Suzhou PerkinElmer Medical Laboratory)] and GBA genetic testing [LR-PCR and sequencing (Suzhou PerkinElmer Medical Laboratory)] excluded Gaucher disease.

The positron emission tomography (PET)/computed tomography (CT)(GE Medical Systems, LLC) scan revealed diffuse fluorodeoxyglucose (FDG) accumulation in the enlarged liver and spleen. The maximum cross-section of the spleen was 146x109 mm, with an upper and lower diameter of 275 mm, and a maximum standard unit value (SUVmax) of 4.7 (Fig. 2A). Mixed bone destruction and slightly active metabolism in the vertebral bodies and sacroiliac bone were also observed. Consequently, a fine needle puncture of the liver tissue guided by B ultrasound (Mindray Resona7, Mindray Co., Ltd.) was performed. However, the patient experienced massive abdominal bleeding after the puncture, leading to an urgent laparotomy for hemostasis. The histopathological analysis of the liver (data not shown) revealed small patches of scattered small lymphocyte infiltration in the portal area and hepatic sinusoids, mixed hyperplasia of T- and B-lymphocytes, and edema and degeneration of liver cells. There was no evidence of lymphoma, leukemia or extramedullary hematopoiesis. The initial diagnosis was suspected to be primary myelofibrosis and the patient was treated with prednisone (40 mg/qd, Shandong Xinhua Pharmaceutical Co., Ltd.) and thalidomide (100 mg/qd, Changzhou Pharmaceutical Co., Ltd.). However, 3 months later, fatigue and bone pain worsened, the liver and spleen became enlarged, hemoglobin levels decreased, and the need for red blood cell transfusions continued. Due to the lack of response to the therapy, a second bone marrow aspiration and biopsy were performed in January, 2022. Erythropoiesis was active and hematopoiesis islands were easily visible in the aspirate (Fig. 1C). Bone marrow biopsy was fixed with 10% formalin at room temperature over 6 h and stained with hematoxylin and eosin (H&E), with a protocol of 5 min of hematoxylin (Beijing Solarbio Science & Technology Co., Ltd.) and 1 min of pure eosin (Beijing Solarbio Science & Technology Co., Ltd.), and subsequently observed under a microscope (Axio Imager A2, Carl Zeiss AG). The biopsy specimen revealed aggravated fibrotic changes, with extensive collagen fibrosis in the bone marrow stroma (Fig. 1D), highlighted by reticulin

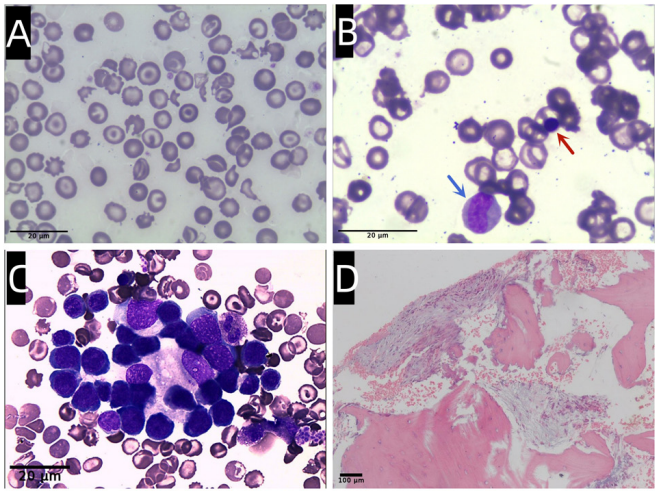

Figure 1. (A) Peripheral blood smear showed target-shaped, elliptic, teardrop shaped, helmet shaped and spherical erythrocytes(magnification, x1000). (B) Peripheral blood smear revealed nucleated red blood cell (red arrow) and metamylocyte (blue arrow) (magnification, x1000). (C) Bone marrow aspirate revealed erythropoiesis was active and hematopoiesis islands were easily observed(magnification, x1000). (D) The bone marrow biopsy specimen exhibited extensive collagen fibrosis and was markedly observed in the stroma (magnification, x100).

stains. Subsequently, ruxolitinib (5 mg/bid, Novartis Pharma Stein AG) was added to the treatment regimen. A repeat of a PET/CT examination indicated multiple bone lytic destruction with increased glucose metabolism and intramedullary hemorrhage, hepatosplenomegaly and internal hemorrhage. The maximum cross-section of the spleen was 164x112 mm, with an upper and lower diameter of ~283 mm, and an SUVmax of 6.3 (Fig. 2B).

Operative and pathological findings. Symptoms of spleen compression were deteriorating. At 6 months after the onset, the patient underwent splenectomy, with the spleen weighing 1,200 g. Nodules and hemorrhage were observed in the spleen specimens (Fig. 3A and B) and were also visible in the liver during the surgery. The splenic parenchyma was barely visible, almost completely replaced by hyperplastic and intensified connective tissue. The splenic specimens were examined using H&E staining 6 min at room temperature and immunohistochemistry according to the protocol. The procedure of immunohistochemical examination was performed. Firstly, a paraffin-embedded section (4-μm-thick) was acquired after fixing a tissue sample by 10% formalin at room temperature over 6 h. The samples were then incubated with 3% H_2O_2 for the inactivation of endogenous peroxidase at room temperature for 15 min. Subsequently, 50 μl primary antibody were added and incubated in a wet box at 37˚C for 90 min. Secondary antibodies were then added and incubated at room temperature for 25 min. Freshly prepared DAB color developer (Beijing Solarbio Science & Technology Co., Ltd.) was diluted x20 for color development. The section samples were final observed under a microscope (Axio Imager A2, Carl Zeiss AG). Various morphological patterns of vascular neoplasms were presented in a background of hemorrhage, along with solid flaky spindle cells (Fig. 4A and B). The Ki67 index was 60% (Fig. 4C). Immunohistochemical results indicated that the neoplastic

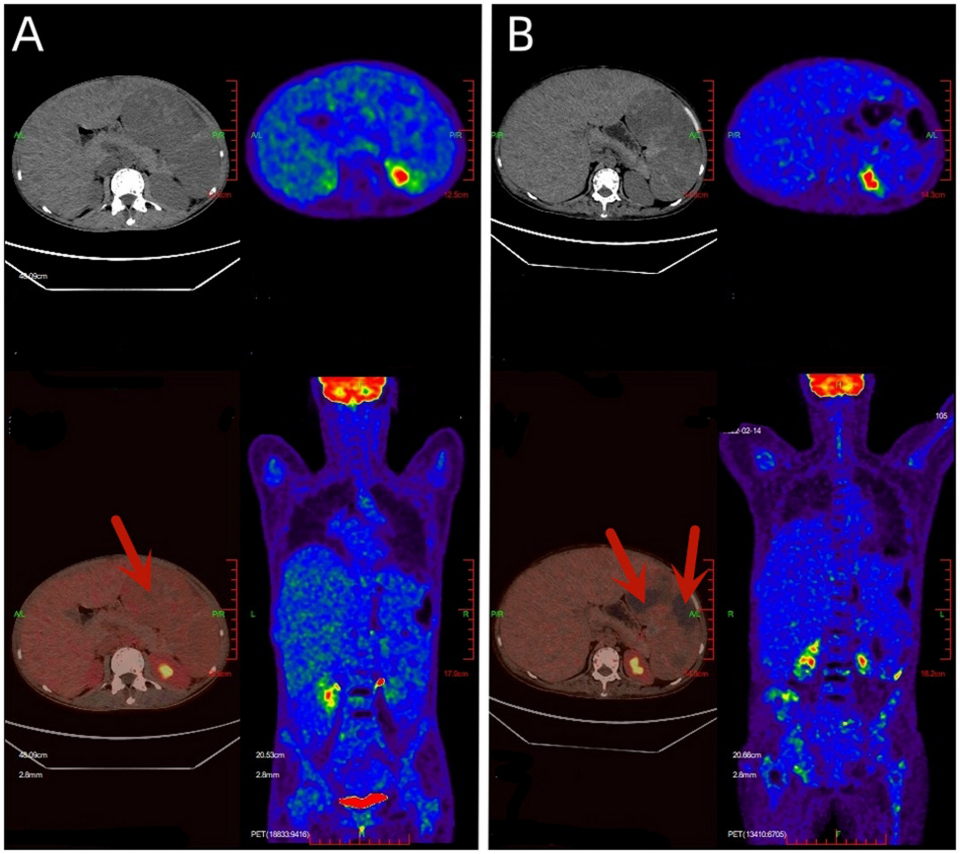

Figure 2. The images in the upper panels represent CT scans, and the images in the lower panels exhibit the fusion of PET and CT. The volume of the spleen increases and multiple necrotic areas without glucose uptake are observed (red arrows). (A) PET/CT performed on September, 2021 revealed maximum cross-section of the spleen was 146x109 mm, with upper and lower diameters of 275 mm, and an SUVmax of 4.7. (B) A PET/CT scan performed on February, 2022 showed maximum cross-section of the spleen was 164x112 mm, with an upper and lower diameter of ~283 mm, and an SUVmax of 6.3. PET, positron emission tomography; CT, computed tomography; SUVmax, maximum standard unit value.

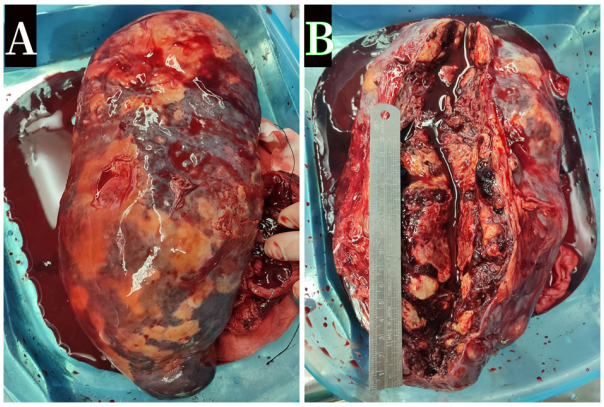

Figure 3. (A) Macroscopic view of the surgical splenic specimen. (B) The cut surface of the excised spleen specimen indicated hemorrhagic and nodular lesions.

cells were positive for CD31, CD34, ERG and Factor VIII (Fig. 4D-G), and negative for human herpesvirus 8 (HHV-8) and CD8 (Fig. 4H and I), supporting an endothelial origin hypothesis. The final diagnosis was splenic angiosarcoma. However, the patient was not suitable for receiving chemotherapy due to her poor medical condition after the surgery. Intermittent red blood cell transfusions were required and the patient passed away 11 months after the surgery.

Discussion

Angiosarcomas, rare soft-tissue sarcomas originating from endothelial cells, often exhibit a high metastatic rate and present with a poor prognosis (1,4,5). They can manifest in various locations throughout the body, with cutaneous presentations being most commonly detected. Prior information on these uncommon tumors has been primarily derived from case series, suggesting that tumor behavior may be influenced by the site of origin (6). Notably, secondary breast angiosarcomas can result from therapeutic radiation or chronic lymphedema (1). By contrast, splenic angiosarcoma is exceedingly rare and highly aggressive, presenting in diverse ways across cases (5,7). Common symptoms include abdominal pain, distension and splenomegaly (8). Additionally, patients may experience anemia, leucopenia, elevated lactate dehydrogenase levels and thrombocytopenia; however, these symptoms are occasionally reported (9). However, these vague presentations can lead to a delayed diagnosis, and such cases may initially resemble blood disorders, characterized by hematological issues and splenomegaly (10). In a subset of cases, splenic rupture occurs as an initial presentation, usually signifying a poor prognosis (9,11-13). Throughout the course of the disease, metastases are common, often occurring early and extensively (14). The liver, lungs, lymph nodes, bone marrow and bone are frequent sites of metastasis (4,10). A previous study

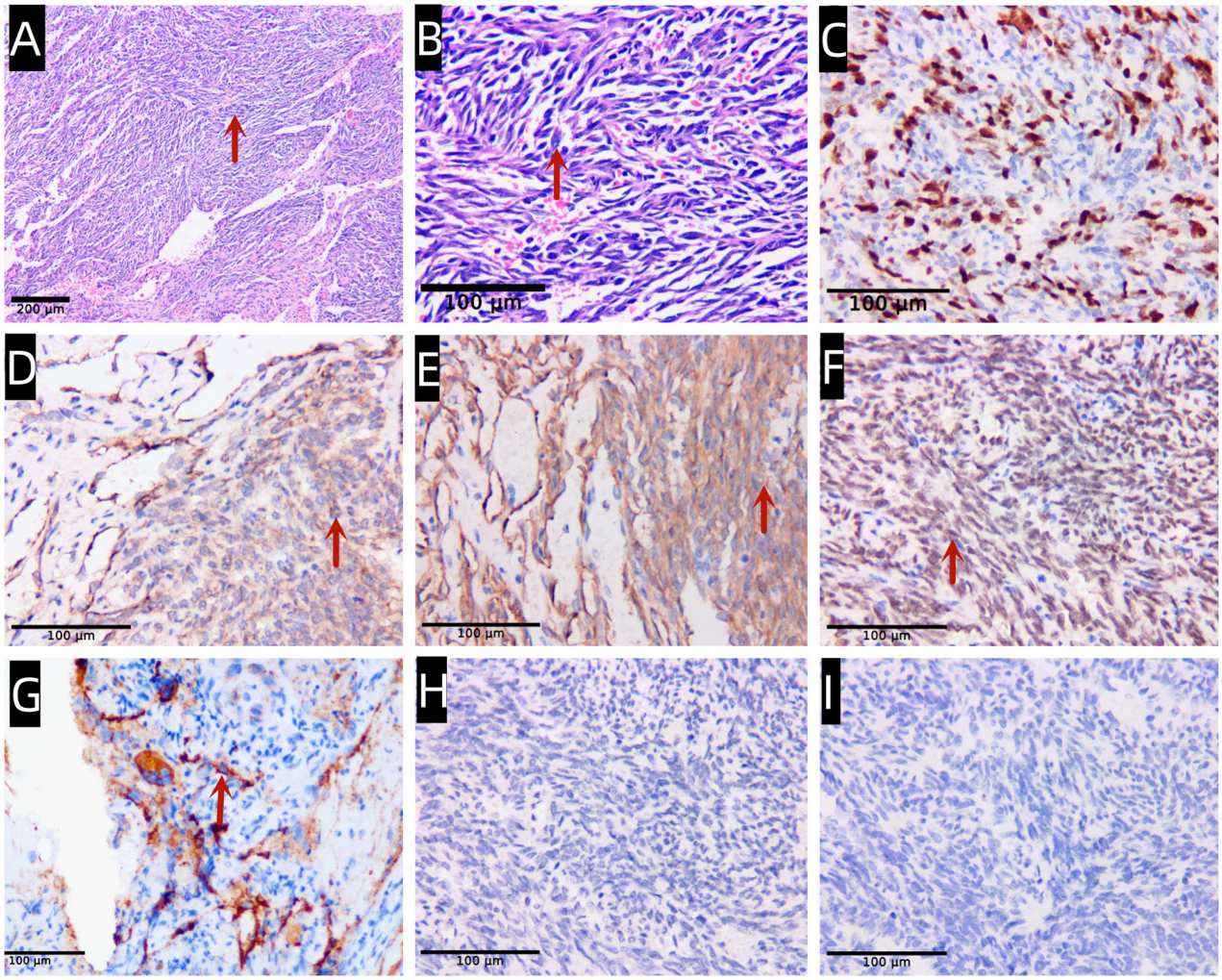

Figure 4. The splenic specimens are examined using H&E staining and immunohistochemistry. (A) Spleen structure was destroyed and almost replaced by a vascular neoplasm with various morphologic patterns in a background of hemorrhage (as indicated by the red arrow; magnification, x100). (B) The splenic histopathology showed hyperplasia of connective tissue and solid flaky spindle-shaped cells were observed (as indicated by the red arrow; magnification, x400). The tumor cells are positive for (C) Ki-67, (D) CD31, (E) CD34, (F) ERG and (G) Factor VIII, negative for (H) HHV8 and (I) CD8. Magnification, x400. The red arrows indicate areas that were typically immunohistochemically positive for tumor cells.

suggested that the median overall survival time of patients with splenic angiosarcoma is ~8.1 months (15). Splenic rupture is an independent indicator of adverse outcomes, while early diagnosis and surgical resection prior to rupture are associated with an improved prognosis (15).

In the present study, a case was reported which initially presented with anemia and bone marrow fibrosis, mimicking primary myelofibrosis, ultimately being diagnosed as splenic angiosarcoma through splenectomy. The patient had no history of prior disease, and no other risk factors were identified. The young woman initially reported fatigue and bone pain. Following admission, anemia, myelofibrosis and splenomegaly were confirmed. The initial diagnosis was that of early myelofibrosis, and the patient received prednisone and thalidomide treatment. However, the symptoms of splenic compression did not alleviate following treatment, and the progression of cachexia was evident. Therefore, the purpose of splenectomy was to relieve the symptoms of splenic compression and define the diagnosis of the primary disease. Surgery was recommended after the multidisciplinary consultation, and the final diagnosis was of splenic angiosarcoma following splenectomy. A previous study reported varying FDG accumulation levels in the primary angiosarcoma of the spleen, with different distribution patterns, including diffuse, peripheral, or multiple nodular type (16). In the case described herein, PET-CT indicated hepatosplenomegaly and internal hemorrhage, accompanied by multiple bone lytic destruction. In the vast majority of previous splenic angiosarcoma case reports, patients initially presented with gastrointestinal symptom or spontaneous splenic rupture (5,13,17). The initial manifestation of anemia and bone marrow fibrosis was a noteworthy observation in the patient in the present case report, since this specific case type may resemble blood diseases with hematological disorders and splenomegaly.

Notably, persistently high ferritin levels were observed in the present case. A previous retrospective study suggested that patients with angiosarcoma with elevated ferritin levels had a significantly poorer overall survival in comparison with patients with normal ferritin levels (18). Additionally, the patient in the present study exhibited a significant decrease in fibrinogen,

which aligns with reports in the literature indicating that patients with angiosarcoma have a higher risk of bleeding, and low plasma fibrinogen levels are predictive markers of a poor prognosis (19,20). In the case reported in the present study, it was hypothesized that excess splenic hemorrhage resulted in substantial fibrinogen consumption, with a subsequent marked reduction in fibrinogen levels. Furthermore, liver invasion may also contribute to reduced fibrinogen synthesis. The present case report highlights the importance of considering bleeding risks for patients with splenic angiosarcoma complicated by hypofibrinogenemia. Therefore, prior to performing a puncture or surgery, it is essential to replenish exogenous fibrinogen to normal levels to prevent hemorrhage.

The accurate diagnosis of splenic angiosarcoma requires a triple assessment, combining clinical examination, imaging findings and pathology (21). Nonetheless, the heterogeneity of clinical presentations and non-specific imaging findings render the definitive diagnosis of splenic angiosarcoma challenging, with histopathological analysis remaining crucial. Splenic angiosarcoma cells typically express multiple markers of vascular differentiation (including CD31, CD34, factor VIII and vascular endothelial growth factor 3) and at least one marker of histiocytic differentiation (CD68 or lysozyme) (4). In the case in the present study, neoplastic cells tested positive for CD31, CD34, ERG and Factor VIII, and negative for HHV8 and CD8. Given the rarity of primary angiosarcoma of the spleen, diagnosis often involves excluding other malignant diseases (8). In the patient described herein, an initial fine needle liver biopsy did not yield a definitive diagnosis, prompting further assessment of its origin. Additional tissue obtained during splenectomy provided additional effective insight into the histological features of a vascular neoplasm, facilitating subsequent immunohistochemical analyses that confirmed the diagnosis of splenic angiosarcoma. Ultimately, a histopathological examination disclosed the diagnosis of primary angiosarcoma of the spleen, with spreading to the liver and bone. The present case report underscores the importance of using a panel of vascular differentiation makers for the confirmation of the diagnosis when encountering unexplained splenomegaly without hematological malignancy.

Given the variability in symptomatology and the potential for life-threatening complications, early diagnosis is crucial. Biopsy acquisition prior to surgery for diagnostic purposes is risky, due to the potential for bleeding and seeding. As a result, histological diagnosis is generally only feasible following splenectomy, which serves both diagnostic and therapeutic purposes, as splenectomy is the treatment of choice for this disease. A previous retrospective review of 145 patients with angiosarcoma demonstrated that primary surgery resulted in an improved overall and progression-free survival (22). Considering the aggressive nature and high mortality rate of the disease, it is worth considering that splenectomy without rupture may significantly extend patient survival. Moreover, PET-CT is able to reveal characteristic changes and detect any metastatic disease (16,23). Metastasis at the time of diagnosis is a common finding and is indicative of a poor prognosis. Therefore, splenectomy, when performed after early diagnosis in the absence of metastatic disease, is associated with a comparably improved prognosis. In the present case, chemotherapy was not a suitable option due to the poor condition of the patient at the time of diagnosis, ultimately passing away 11 months after the surgery. Consequently, it is suggested by the authors that splenectomy should be considered both for diagnosis and treatment when appropriate in cases with a high suspicion of primary splenic angiosarcoma.

The exact pathogenic mechanisms underlying angiosarcoma remain incompletely understood. Yamamoto *et al* (24) reported that tumor cell-derived stem cell factor may potentially influence the increased presence of mast cells, which in turn, could contribute to the proliferation of tumor cells, ultimately driving the progression of angiosarcoma. Primary myelofibrosis usually occurs in elderly individuals, with a median age of onset of 60 years. Additionally, mutations in *JAK2*, *MPL* and *CALR* genes have been identified in the majority of primary myelofibrosis, and patients negative for these three mutations are relatively rare (25). In this present study, the patient was 35 years of age without driver mutations, thus being definitely diagnosed with splenic angiosarcoma. Therefore, it was speculated that the myelofibrosis in this patient was most likely secondary to angiosarcoma. It is conceivable that the anemia may have resulted from hypersplenism or extensive secondary myelofibrosis, triggered by angiosarcoma. In addition, abnormally elevated peripheral blood nucleated red cells were observed. The destruction of red blood cells was considered, due to intrasplenic hemorrhage, compensation for erythroid proliferation in the bone marrow, and extramedullary hematopoiesis as possible causes of this type of abnormally increased numbers of nucleated erythrocytes.

In conclusion, the present study describes the case of a patient in whom the initial symptoms were suggestive of anemia and bone marrow fibrosis, mimicking primary myelofibrosis. However, this was eventually attributed to splenic angiosarcoma. The present case underscores the importance of considering splenectomy for the acquisition of histopathological evidence, further highlighting the value of employing a panel of markers for vascular differentiation, in order to aid in the diagnosis of angiosarcoma.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Authors' contributions

All authors (MW, ZL, LL, WZ and JL) contributed to the conception and design of the study. Data collection was performed by MW and ZL. Data analysis was performed by LL and WZ. The first draft of the manuscript was written by MW and JL. ZL, LL and WZ confirm the authenticity of all

the raw data. All authors have read and approved the final version of the manuscript.

Ethics approval and consent to participate

Written informed consent was obtained from the patient described in the present case report.

Patient consent for publication

Written informed consent was obtained from the patient for the publication of personal information-related data and any related images.

Competing interests

The authors declare that they have no competing interests.

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