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Thrombocytopenia as first manifestation of splenic angiosarcoma

Simon Raffel · Bert Hildebrandt · Christian Grieser · Stefan Pahl · Isrid Sturm

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Dear Editor,

Angiosarcoma is a rare mesenchymal neoplasia with an incidence of only 0.14–0.25 cases per million [1]. Most common sites of occurrence are skin and superficial soft tissues, followed by breast, liver, spleen, and bone. Early stage metastases to lung, liver, and lymph nodes are common and overall prognosis is poor [2, 3]. Only approx. 5% of angiosarcomas originate from the spleen, and the existing casuistic reports suggest a dismal clinical prognosis.

Here, we describe a rare case of splenic angiosarcoma leading to thrombocytopenia as first manifestation of the disease.

A 64-year-old woman presented as hematologic outpatient in March 2008 with isolated thrombocytopenia (58/nl) and chronic, slowly progressive fatigue. Thrombocytopenia was first noted in 2006. Pre-existing medical conditions and physical examination were unremarkable. The remaining full blood count was without pathological findings (hemoglobin 12.2 g/dl, normal erythrocyte indices, leukocytes 6.9/nl with normal differential count). Differential diagnosis for moderate thrombocytopenia included various causes for

immune destruction of platelets, impaired thrombocyte production, and hypersplenism. However, laboratory screening tests for lymphoma, autoimmune diseases, and virus infections resulted negative.

A bone marrow aspirate gave an unspecific picture with unremarkable cellular properties, normal count of megakaryocytes but diffuse infiltration of mature plasma cells up to 15%. A bone marrow biopsy showed hyperplastic left shifted granulopoiesis, left shifted megakaryopoiesis, and erythropoiesis in a hypercellular tissue, partially accompanied with B-cell-rich non-monoclonal lymphocytic aggregates.

An abdominal ultrasound showed massive splenomegaly with multiple highly echogenic circular lesions. Multislice computed tomography (MSCT) confirmed splenomegaly (measuring 21 cm craniocaudal) and revealed multiple hypodense lesions with central hyperdense structures (Fig. 1a). In addition, multiple lytic bone lesions in the vertebral column were present. MSCT-guided biopsy of these lytic bone lesions showed infiltrates of a mesenchymal neoplasia containing atypic mitotic bodies, a proliferation fraction of 40% (MIB-1) and negativity for pancytokeratin and S 100. Immunohistochemistry stained positive for vimentin, CD31, CD34, and factor VIII, but negative for desmin, actin, and HHV8, leading to the diagnosis of angiosarcoma (Fig. 2).

Meanwhile, the patient suffered from symptomatic splenomegaly (abdominal pain, symptoms of GI tract displacement), and since splenic angiosarcoma may be complicated by splenic rupture [4, 5], an elective splenectomy was performed. Histological examination confirmed the diagnosis of primary splenic angiosarcoma. In addition, the patient was offered palliative chemotherapy and infusions of bisphosphonates; the latter were started immediately. Chemotherapy with paclitaxel (90 mg/m² weekly) began with a delay of several weeks (patient's choice) at a

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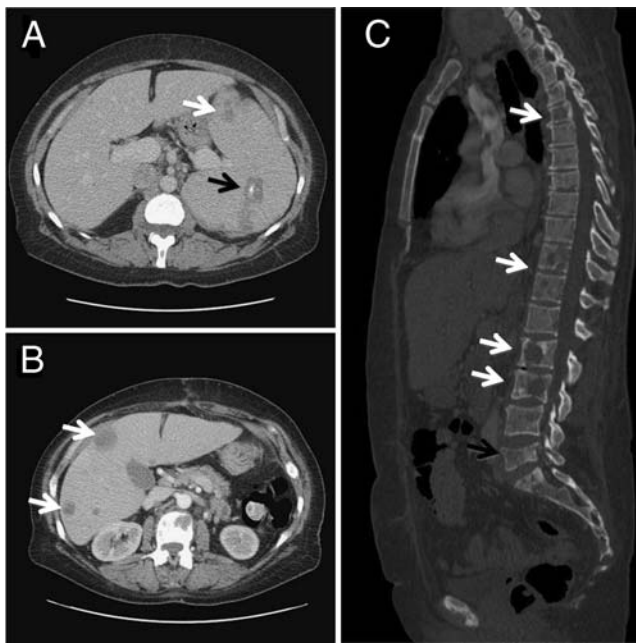


Fig. 1 Initial MSCT examination presented splenomegaly with multiple hypodense lesions (**a**; *white arrow*) which were partially calcified (**a**; *black arrow*); MSCT after splenectomy (3 months later) showed multiple new liver metastases (**b**; *white arrows*); furthermore, sagittal reformation revealed diffuse progression of the lytic bone lesions now present in all parts of the axial skeleton (**c**; *white arrows*) and an old fracture of the fifth lumbar vertebrae (**c**; *black arrow*)

time when massive vertebral pain was already present. Following MSCT examination (3 months after the initial study), the patient presented a progressive disease with diffuse metastases to multiple vertebral bones (Fig. 1b) and new liver metastases (Fig. 1c). Meanwhile, the blood count was leukerythroblastic with leukocyte counts up to 63/nl resulting from advanced bone marrow infiltration. Thrombocyte counts further decreased to a minimum of 10/nl, and progressive anemia required multiple consecutive transfusions of packed RBC. Approximately 4 months after diagnosis, the patient died from diffuse gastrointestinal bleeding.

Primary splenic angiosarcoma is a very rare malignancy with a known aggressive behavior. Leading symptoms are usually abdominal pain and hematologic abnormalities; thrombocytopenia has been described in 33% of cases [6]. Metastases are common, mainly in liver and bone.

We present this case to emphasize that splenic angiosarcoma, although rare, should be considered in the differential diagnosis in patients with hematologic abnormalities of unexplained origin and parenchymal lesions of the spleen.

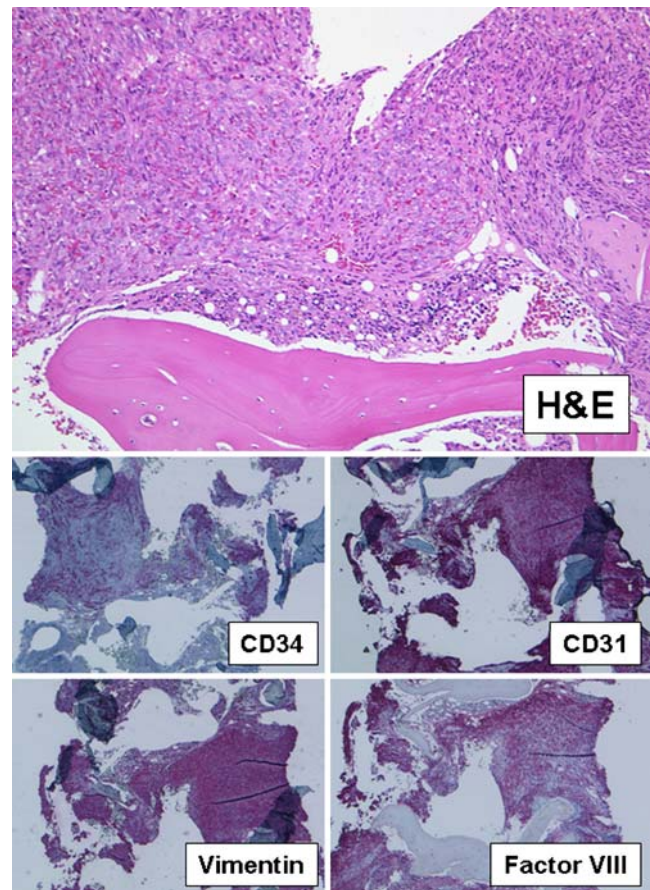


Fig. 2 Histological analysis of MSCT-guided biopsy of lytic bone lesions in the vertebral column showed a mesenchymal neoplasia containing atypic mitotic bodies infiltrating spongy bone tissue (upper part, H&E, $\times 100$). Immunohistochemical stainings were positive for CD31, CD 34, factor VIII, and vimentin (lower panels, immunoperoxidase, $\times 40$) leading to the diagnosis of angiosarcoma

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