Primary splenic angiosarcoma: a very rare and aggressive neoplasm with a poor prognosis

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CONFLICT OF INTEREST None declared
A 78-year-old man was admitted to hospital on account of persistent pain in the left part of the abdomen and under the left scapula. On physical examination tenderness on palpation in the left epigastric region was observed. Laboratory tests showed: normocytic anemia, red blood cells (RBC) – $3.72 \times 10^6$ ul, thrombocytopenia (90x10^3 ul), an increased activity of alkaline phosphatase (ALP) – 277 U/l, D-dimers - 5055 ng/mL, C reactive-protein (CRP) - 23.09 mg/l and elevated erythrocyte sedimentation rate (ESR) – 160 mm/h. First abdominal ultrasound showed an enlargement of the spleen (size: 150x87mm) with a focal angioma-like lesion in its lower region and in the central segment of the spleen, another area with an increased echogenicity, 45x46mm, surrounded by a thoroughly encapsulated surface that may indicate a hematoma-type lesion around the angioma. An abdominal computed tomography (CT) scan revealed an enlarged spleen (up to 145 mm in the longest axis) with multiple heterogeneous focal changes of different size affecting a significant part of the splenic parenchyma and remarkably distorting its outer outline (Figure 1 A, B). Due to persisting pain in the left subcostal another abdominal ultrasound was performed (after 16 days following the first examination); it showed that the angioma-like lesion in the central region of the spleen grew up to the size of 81x54x68mm. Patient was transferred to the Department of General and Vascular Surgery, where he was qualified for splenectomy. The histopathological examination of the resected spleen, microscope image and immunohistochemical profile (CD34+, CD31+, CK, AE1/AE3, Ki67 about 60% - in the fields of the highest activity) indicated splenic angiosarcoma [Figure 1 E, F, G, H]. Three weeks after being diagnosed, a CT scan of the chest, abdomen and pelvis showed numerous small liver metastatic lesions and an infiltration of the gastric and peritoneal region (Figure 1 C, D).

Primary splenic angiosarcoma is a very rare malignant neoplasm arising from splenic endothelial cells. The annual incidence rate of the neoplasm is between 0.14 and 0.25 of cases per one million with the peak incidence in patients aged 50-60 years [1,2]. Pathogenesis of
this type of neoplasm still remains unknown. Some authors believe that angiosarcoma grows out of such benign lesions as hemangioma and hemangioendothelioma. There is also some data on impact of toxic substances such as thorium, arsenic and vinyl chloride on angiosarcoma development [2]. Splenic angiosarcoma does not cause any typical clinical signs, the only symptom occurring in over 80% of patients is nonspecific pain in the left epigastric area. Splenic rupture is sometimes the only manifestation of splenic angiosarcoma [3]. Metastasis index is from 69% to 100 %, and the main organs to which the disease spreads are the liver (89%), lungs (78%), lymphatic nodes (56%) and bones (22). Like most angiosarcoma, surgical resection is the main treatment, early adjuvant chemotherapy only theoretically inhibits the metastatic potential of angiosarcoma [4]. The average survival rate among patients suffering from the disease is 12 months following the final diagnosis [5]. Splenic angiosarcoma should be suspected in each patient with atypical pain in the left subcostal area, an enlargement of the spleen with atypical focal changes, thrombocytopenia and anemia of unknown etiology.

BIBLIOGRAPHY


Figure 1  A- abdominal computed tomography scan showing multiple irregular heterogeneous masses in the spleen coronal scan; B- transverse scan
Figure 1  C- abdominal computed tomography scan after splenectomy with numerous liver metastases coronal scan; D- transverse scan
Figure 1  Microscopic view of the splenic angiosarcoma. E- clusters of spindle tumor cells infiltrating the spleen parenchyma, with atypical blood vessels (magnification × 200).

Immunohistochemical examination of the splenic specimen. F- specimen stained positive for CD31 (magnification × 200); G- specimen stained positive for CD34 (magnification × 200); 

H- Ki-67 proliferation index less than 60% (magnification × 200)