1130-0108/2008/100/6/373-379
Revista Española de Enfermedades Digestivas

Rev Esp Enferm Dig (Madrid)
Vol. 100, N.° 6, pp. 373-379, 2008

Letters to Editor

Primary angiosarcoma of the spleen

Key words: Angiosarcoma. Spleen.

Dear Editor,

Primary angiosarcoma of the spleen is a rare tumor, with no more than 200 cases reported in the literature worldwide and with a poor prognosis. The symptoms and radiologic findings are non specific.

We present a new case of primary angiosarcoma of the spleen that confirms his aggressiveness. We report a case of a 63-year-old man with arterial hypertension and prostatic syndrome, who was admitted to our hospital with a history of 3 weeks of epigastric abdominal pain, sometimes radiating to the left hypochondrium, accompanied of sensation of fullness but not related to the ingestion and slimming of 3 kg of weight.

The physical examination revealed slight pain in the left hypochondrium with normal bowel sounds and moderated splenomegaly.

Laboratory examinations revealed: haemoglobin 11 g/dL, total white blood cell 12.98 x 10^9/L, platelets 520 x 10^9/L. Normal coagulation. Biochemistry: creatinine 1.4 mg/dL, other parameters are normal. Tumor markers: CA-19.9, carcinoembryonic antigen (CEA), PSA and alpha-fetoprotein (AFP) were normal.

Abdominal CT scan showed a moderate heterogeneous splenomegaly with multiple areas with a low-density signal compatible with necrosis and other nodular areas suggestive of infiltrative pathology, without lymphatic ganglions, and a liver cystic of 2 cm (Fig. 1). The splenic biopsy by CT scan showed a hematic smear with few cells and isolated nuclear atypia does not diagnose of malice.

Splenectomy was performed. Histological examination revealed a splenic angiosarcoma. The spleen weighted 658 g and measured 18 x 11 x 7 cm. Tumor cells were positive for CD31 and CD34 (vascular markers) (Fig. 2).

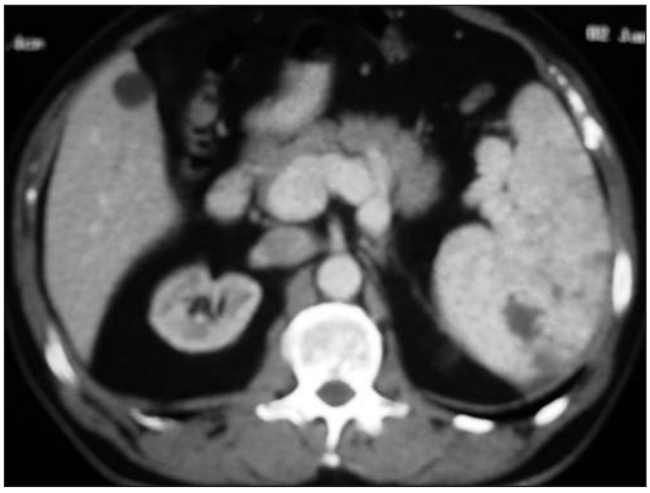

Fig. 1. Moderate splenomegaly with nodular areas suggestive of infiltrative pathology.

One month before the surgery a bone gammagraphy showed an anomalous increase of the tracer in the fifth right rib and sacredly suggestive of bone metastases, and abdominal CT scan showed liver metastases. The patient died 2 months after the surgery.

Primary angiosarcoma of the spleen was describe by Langhans in 1879 and is a very rare tumor that represents the 1-2% of the sarcomas.

The presenting symptoms are non specific and variable, but abdominal pain in the left hypochondrium, weight loss, anemia and splenomegaly are frequently present (1-3). Hemoperitoneum due to splenic rupture is seen in up to 30% of cases and often is the first manifestation of the disease (4).

It is in the habit of presenting in the middle ages of the life but also there are described cases in children (5). Radiologic findings are non specific, but the diagnosis should be suspected in a patient who presents with splenomegaly but without evi-

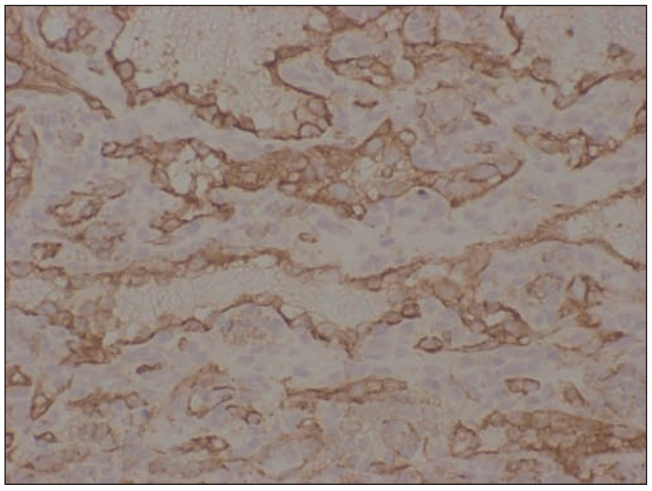

Fig. 2. Immunocystochemical marker CD31+.

dence of lymphoma, leukemia, malaria or portal hypertension (6). Metastases are present in 69-100% of the cases. Liver (41-89%), lung (78%), bone or bone marrow (2-44%) and lymphatic ganglions (3-56%) are the most common sites (2,5).

Splenectomy is the recommended treatment. Immunocystochemical markers like CD31, CD34, FVRAg, VEGFR3, CD68 and/or lysozime are very specific but frequently not analyzed before the surgery (2,7).

Diagnosis can only be made at histological examination of the splenectomy specimen. Histologically, the tumor is characterized by neoplastic proliferation with diffuse or focal areas of a vasoformative component with cavernous and arborizing channels. The differential diagnosis includes hemangioendothelioma that presents a solid supporter with few blood vessels, scanty atypia and without mitosis, and the coastal angioma that presents vascular channels without atypia and pleomorphic tumor cells (7).

Prognosis is very poor and the median survival time is 10-14 months. A minory are still alive after 5 years (2,3). The adjuvant treatment with chemotherapy or radiotherapy have not improved the prognosis of the patients (2-4).

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