Primary splenic angiosarcoma with multiple osseous metastases: a rare presentation of the neoplasm

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Abstract Primary splenic angiosarcoma is very rare malignant neoplasm and it is extremely rare to find splenic angiosarcoma with osseous metastases. This study reported a 53-year-old male patient with massive metastases in almost whole vertebrae. Splenectomy, chemotherapy and radiotherapy were performed. The patient survived 6 months after diagnosis.

Key words spleen; angiosarcoma; osseous metastases

First described in 1879 by Langhans [1], splenic angiosarcoma is a rare malignant tumor with very poor prognosis. Up to now, only around 200 cases are reported around the world [2]. Splenic angiosarcoma is highly invasive with a high propensity for hepatic metastases. However, osseous metastasis is extremely rare. Here we reported such a case.

Case report

In July 2012, a 53-years-old male patient presented to his primary care physician with lumbago without fever, shivering, nausea, abdominal distension. Chest computed tomography (CT) demonstrated that irregular hypo dense shadow in enlarged spleen and imaging of chronic bronchitis and multiple destructions of sternum and thoracic vertebra. Bone emission CT demonstrated high radioactive concentration in the area of left 3rd rib and a mass of radioactive enhanced shadow on upper-left abdomen, which was highly suspected to be the spleen. He did not receive treatment at that time.

In August 2012, the patient’s contrast enhanced CT scanning demonstrated large spleen with heterogeneous lesions and mixed hypo density in arterial phase; the foci presented isodense with small enhanced patches in delay and venous phase. The maximum cross-section area of the lesion was 114 mm × 121 mm (Fig. 1).

Magnetic resonance imaging (MRI) presented massive metastases in cervical vertebra-7, thoracic vertebra 1–12, lumbar vertebra 1–5, sacrum 1 and 3 and pelvis (Fig. 2).

All tumor markers such as AFP, CEA, CA19–9 and CA12–5 were negative.

The patient soon underwent splenectomy. The surgeon observed an enlarged spleen with a 10 cm × 10 cm tumor capsule and a 3 cm × 3 cm lymph node could be touched near spenic hilum.

Under the macroscopy the tumor was composed of atypical vascular neoplasm of anastomosing vascular channels lined by endothelial cells (Fig. 3). In some area, the neoplastic cells grouped in shape of nest, rope and blocks (Fig. 4), within them, primary vessels were detectable and immunohistochemistry staining showed that Vim(+), CD8(–), SMA(–), PCK(–), WT-1(–), Ki-67(+10%), CK5/6(–), CD31(+), CD34 local(+), factor XIII(–), CD68(–), LYS(–), PAS(–) (Fig. 5, 6).

Based on these examinations, the diagnosis of angiosarcoma was made.

Since the laboratory revealed mild anemia and there were multiple osseous metastases, chemotherapy and multiple-area radiotherapy would be intolerable immediately after the surgery. So, the patient was managed with immunopotentiation therapy for a month.

Adriamycin and ifosfamide regimen started a month later. After 2 cycles of treatment, MRI manifested the lesion expanded. The patient’s chemotherapy regimen was switched to dacarbazine and cisplatin for 1 cycle and he refused further chemotherapy treatment. The patient died in Jan 2013, 6 months after the diagnosis.

Discussion

Spleenic angiosarcoma is extremely rare, very aggres-
sive and with high metastatic rate. It originated from malignant hyperplasia of splenic vascular endothelium.

No etiologic association has been clearly established, but caustic factors may include radiation, chronic lymphoedema, exposure to toxins and familial syndromes[3].

In our case, the patient claimed non-involvement in any of the above circumstances. His main complains was lumbago. Laboratory showed anemia and thrombocytopenia.

Since there is no specific tumor marker for splenic angiosarcoma and it is unidentifiable in both serological and immunohistochemical examinations, diagnosis of splenic angiosarcoma depends mainly on biopsy and pathological examination.

Typical radiological features of splenic angiosarcoma include: enlarged spleen with ill-defined multiple heterogeneous density.

Fig. 1 An enlarged spleen with a heterogeneous density

Fig. 2 Massive metastases in almost whole vertebras

Fig. 3 HE staining × 100
Fig. 4 HE staining × 200
Fig. 5 CD31 immunohistochemical staining × 400
Fig. 6 CD34 immunohistochemical staining × 400
geneous hypodense masses or with low mixed densities; on arterial phase, the lesion shows less enhancement than parenchyma; on delay phase, the lesion presents with isodense enhancement higher than parenchyma. In the event of large lesion, due to the uneven distribution of blood, the lesion shows variegated enhancements, sometimes affected by fibrosis scar, presents as dotted blocks.

On ultrasound, a complex mass with heterogeneous echotexture is the most common finding. Areas of necrosis and hemorrhage are frequently noted as cystic areas within the mass.

In our case, since the contrast enhance CT meet the typical appearance, associated with the laboratory findings of anemia and thrombocytopenia, the pre-operation diagnosis highly suspected it was splenic angiosarcoma.

In most cases, diagnoses of splenic angiosarcoma through radiologic images are highly challenging, because they mimic images of other vascular splenic tumors, such as hemangiomas, littoral cell angiomas, lymphangiomas, hemangiopericytomas, and epithelioid vascular tumors.

Splenectomy, combined chemotherapy and radiology are standard treatment for splenic angiosarcoma, however, due to it extreme malignance, high invasive and fast proliferation, all treatments are rarely curative. Optimal therapies are still in searching, though there are literatures saying that paclitaxel, gemcitabine, doxorubicin, ifosfamide, dacarbazine, bevacizumab, etoposide and cisplatin may bring clinical benefits.

The most common metastatic sites include liver, lung, lymph node, bone, bone marrow, and soft tissue.

In our case, although the patient underwent a R0 splenectomy with microscopically negative margins, the tumor developed so very fast that there wasn’t enough time to locate the sensitive chemotherapy medicine. At the same time, radiology incurred severe side-effects. The patient died of severe anemia 6 months after diagnosis.

References