CASE REPORT

ANGIOSARCOMA OF THE SPLEEN

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Abstract

A thirty seven year old previously healthy lady presented with episodes of vague left sided upper abdominal pain of three months duration. Investigations revealed anaemia and thrombocytopenia. The ultrasound scan and the computed tomography of the abdomen showed splenomegaly with multiple echogenic nodules suggestive of a lymphoproliferative disease. A splenectomy was done. Multiple hemorrhagic liver nodules were identified during the surgery. The histology revealed a well differentiated angiosarcoma of the spleen with secondary deposits in the liver. Primary angiosarcoma of the spleen is a rare mesenchymal malignant tumor of vascular origin and is a rare cause of hypersplenism due to sequestration of blood in the spleen.

Introduction

Hypersplenism occurs due to sequestration of blood in the spleen and increased phagocytosis of the cellular components of blood. Angiosarcoma of the spleen is a rare mesenchymal malignant tumour of vascular endothelial origin and is a rare cause of hypersplenism. The most common neoplasms of the spleen are of haematolymphoid origin, e.g.; secondary involvement or primary lymphomas.

The non haematolymphoid splenic tumours are rare, and of them the commonest is angiosarcoma, with an annual incidence of 0.14–0.25 per million[1-3]. The mean age of presentation ranges from 50 to 79 years[1,3]. This disease was first reported in 1879 with only 200 cases reported since then in the literature[3].

Case Presentation

A thirty seven year old lady presented with episodes of vague left sided upper abdominal pain of three months duration. The past medical history was unremarkable. On examination, she was pale. Her abdomen was distended with massive splenomegaly. The liver was not palpable. Investigations revealed anaemia (hemoglobin 8.2 g/dl) and thrombocytopenia (platelets 97 × 10^9/l). Computerized tomography of the abdomen showed splenomegaly with multiple echogenic nodules suggestive of a lymphoproliferative disease. The liver was mildly enlarged and no focal lesions were seen. A laparotomy was performed and splenectomy was done. At laparotomy, multiple haemorrhagic liver nodules were observed. The cut surface of the spleen revealed multiple pale irregular solid lesions. Microscopy of these pale lesions showed extensive coagulative necrosis. The

surrounding tissue showed a tumour composed of numerous, complex, well-formed capillary size vascular channels (**Fig. 1**) lined by cells containing mild to moderately pleomorphic nuclei. The neoplastic cells were strongly positive for CD31 and CD34 (**Fig. 2**). Approximately 5% of the neoplastic cells expressed proliferation marker Ki-67.

The diagnosis of a well differentiated angiosarcoma of the spleen was made. Histology of the liver nodules showed a spindle cell neoplasm with morphological features of vascular histogenesis, which was confirmed by vascular markers, CD31 and CD34. CD 117 and pan cytokeratin were negative. These features were suggestive of secondary deposits from an angiosarcoma.

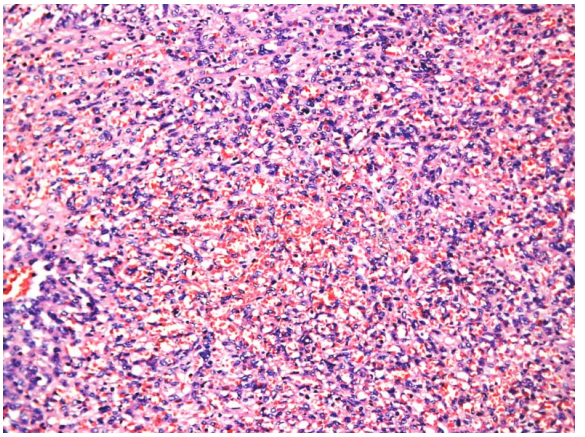
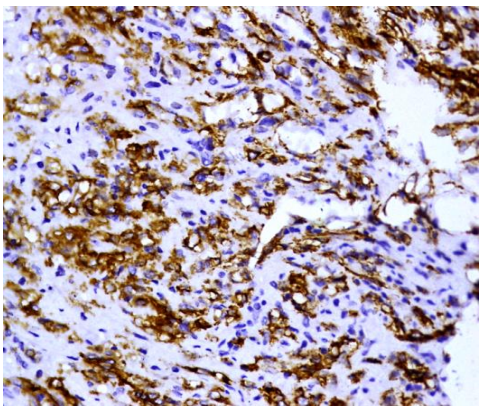


Figure 1 - The splenic tumour showing numerous complex capillary size vascular channels. (Haematoxylin and eosin x200)

CD 34



CD31

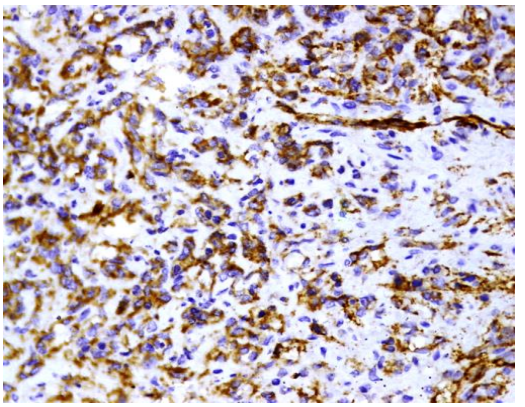


Figure 2 – Splenic tumour showing strong membrane and cytoplasmic positivity for CD34 and CD31. (Dako Monoclonal Mouse Anti-Human, EnVisionTMFLEX system x 400)

Discussion

Hypersplenism is associated with splenomegaly and the common causes include thalassaemia major, malignant lymphoma and myelofibrosis. Vascular neoplasms of the spleen have been reported as rare causes of hypersplenism, and primary angiosarcoma of the spleen is an extremely rare cause of hypersplenism. In one series about 15% of patients with splenic angiosarcoma had thrombocytopenia[5]. Our patient had thrombocytopenia and anaemia.

Primary angiosarcoma of the spleen is a rare and aggressive malignancy that often

harbours metastatic disease at the time of diagnosis and carries a dismal prognosis[3]. There are no specific clinical diagnostic guidelines for this rare entity[1]. Imaging is helpful for the differential diagnosis. The most common ultrasonography findings are splenomegaly and ill-defined solid and cystic masses in the spleen with heterogeneous echo texture. Pre-operative CT imaging is invaluable both for diagnosis and acute assessment of complications[3].

Definitive pre-operative diagnosis of splenic angiosarcoma remains challenging. Biopsy is hazardous due to risk of bleeding and malignant seeding[3]. Histological diagnosis is therefore usually possible only after splenectomy, which is both diagnostic and therapeutic[3].

Metastasis is common at diagnosis indicating poor prognosis. Metastases occur in 69% to 100% of cases of splenic angiosarcoma with 89% involving the liver[1,3]. Irrespective of treatment the median survival remains dismal at 5 months[2,3]. Nevertheless, splenectomy after early diagnosis in the absence of metastatic disease has been shown to have a much better prognosis[2,4]. The presence of liver metastasis at the time of diagnosis in our patient indicates poor prognosis and poor survival time.

Conclusion

Clinical and radiological diagnosis of splenic angiosarcoma are challenging. Definitive histological diagnosis is usually be made after splenectomy since biopsy is contraindicated.

Acknowledgement

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