

Case Report

Primary splenic angiosarcoma: a diagnostic enigma

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ABSTRACT

Primary splenic angiosarcomas (PSA) arise from splenic endothelium are rare and impose a diagnostic challenge preoperative. They can present as asymptomatic splenomegaly however; the commonest presentation is abdominal pain. The spleen can rapidly increase in size and can manifest as spontaneous rupture which would cause peritoneal dissemination of disease. Early metastasis of PSA is seen in liver, lungs, lymph nodes and gastrointestinal system. Preoperative diagnosis requires a high index of suspicion and ultrasound, contrast enhanced computerized tomography may essential to differentiate from splenic hemangioma. Splenic angiosarcoma are best treated with splenectomy with a limited disease, with care taken not to rupture and cause spillage. PSA are resistant to adjuvant radiation and chemotherapy. Mortality is high with median survival rate of only 5 months, irrespective of treatment and hence the need to diagnose before complications. Bisphosphonates, adjuvant radiation with chemotherapy have been attempted to increase disease free survival. We report a case of PSA emphasizes on early preoperative diagnosis to avoid progression of the disease.

Keywords: Primary splenic angiosarcoma, Mesenchymal splenic tumor, Splenic angiosarcoma, Vascular tumor of spleen

INTRODUCTION

Soft tissue sarcomas are tumors that arise in mesenchymal tissues. Angiosarcomas originate from endothelial cells and constitute less than 1% of all cases of soft tissue sarcomas.¹ Primary splenic angiosarcomas (PSA) arise from splenic sinusoidal vascular endothelium and are among the rare malignant tumors with annual incidence of 0.14 to 0.25 cases per million persons.^{2,3} PSA was reported first by Langhans in 1879.⁴ The most common presentation is abdominal pain and about 30% patients present with nontraumatic splenic rupture resulting in peritoneal and vascular dissemination which has worst prognosis.³ Splenectomy before rupture of spleen should be aimed to increase disease free survival.⁵ Irrespective of treatment, PSA has very guarded

prognosis because of difficulties in diagnosis and its extremely aggressive behavior.

We report a case of primary splenic angiosarcoma diagnosed on histopathology after splenectomy for spontaneous rupture with hemoperitoneum, the preoperative diagnosis being hemangioma of the spleen.

CASE REPORT

A 55 years old lady having no co-morbidities presented with history of left upper abdominal pain since one month. She denied any past medical or surgical illness. She had no history of exposure to chemicals as vinyl chloride or any radiation. Her all routine laboratory investigations were normal. Her ultrasound of abdomen revealed splenomegaly with large mass with decreased

echogenicity. Contrast enhanced computed tomography (CT scan) of abdomen pelvis confirmed enlarged spleen with mass of 6 centimeter in diameter having hypervascular rim and low attenuation in the center indicating central necrosis and reported as splenic hemangioma (Figure 1 and 2). Patient was being investigated when she presented in emergency department with severe generalized abdominal pain and distension without history of trauma after 3 days. She was pale having tachycardia and hypotension. She was resuscitated with 2 units of blood and ultrasound abdomen was done which revealed free fluid in the abdomen with internal echoes and ruptured spleen. Paracentesis of the abdomen was sanguineous. She was subjected for emergency exploration. There was about 2 liters of hemoperitoneum with ruptured enlarged spleen with a firm mass near hilum which was presumed as hemangioma. Splenectomy was done (Figure 3). Postoperative course was uneventful and patient was discharged on day 7 after giving vaccination so as to prevent post splenectomy infections.

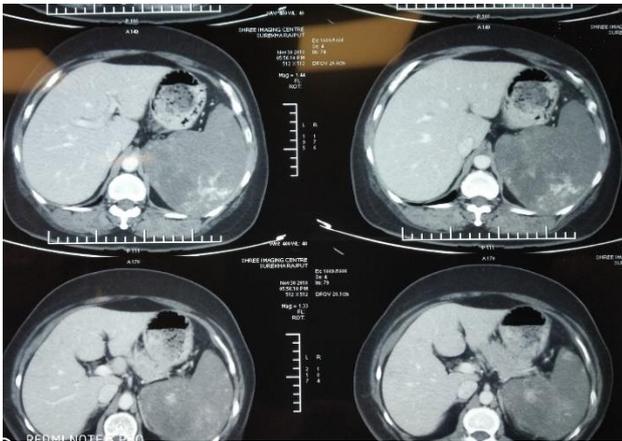


Figure 1: Computerized scan of the abdomen showed mass, with hypervascularity in the spleen suggestive as hemangioma.



Figure 2: Coronal section of the CT scan of the abdomen showing splenomegaly with hyper vascular channels in centre.



Figure 3: Specimen of rupture spleen showing mass near the hilum.

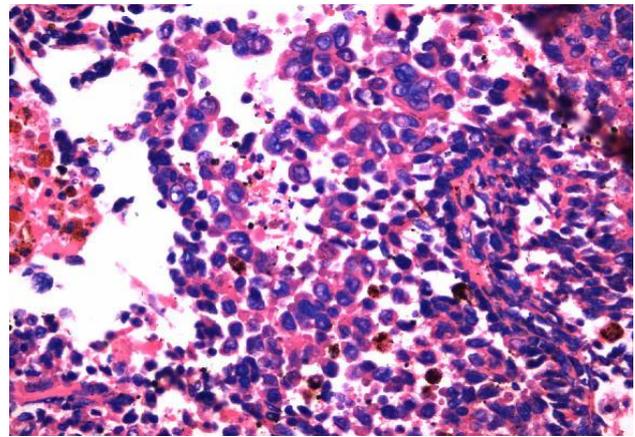


Figure 4: Epithelioid morphology of the tumor HE X400.

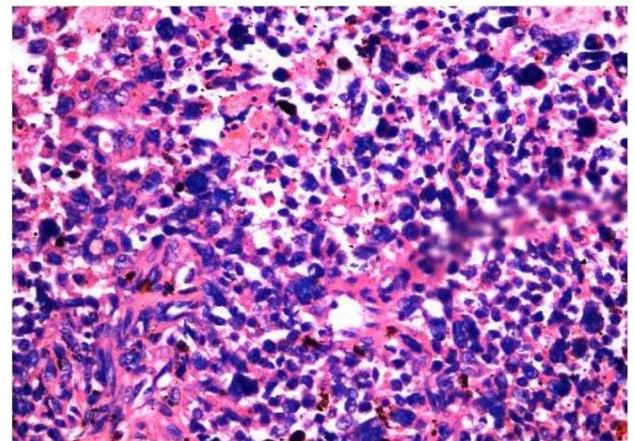


Figure 5: Intracytoplasmic RBC HE X400.

Patient followed up on 12th postoperative day with microscopic histopathology showing cavernous vasoactive pattern with mesenchymal cells showing nuclear atypia and occasional mitosis consistent with neoplastic nature (Figure 4-6) on immunohistochemistry, tumor cells were positive for CD 31, CD 34 and factor VIII related antigen. To our surprise final histopathology report was splenic angiosarcoma. Tumor markers like CA

19-9, CEA and CA 125 were normal. Fluorodeoxyglucose (FDG) positron emission tomography (PET) scan did not reveal any positive activity. Patient was referred to medical oncologist for chemoradiation. Patient was asymptomatic at 3 months follow up, however developed liver metastasis in 5th post-operative month and succumbed to it.

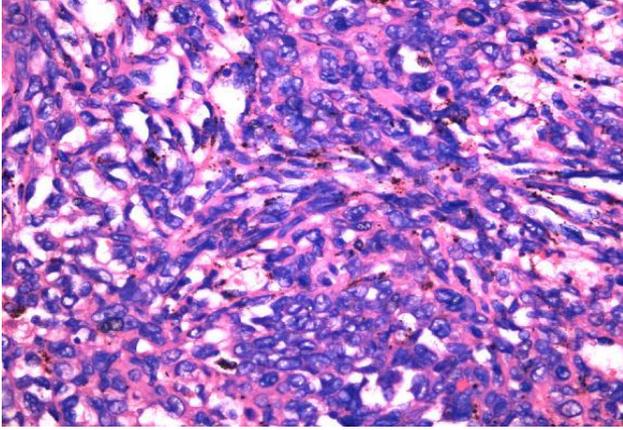


Figure 6: High grade sarcoma HE X400.

DISCUSSION

PSA is a rare and aggressive malignancy arising from vascular endothelium of spleen. It affects most commonly after 40 years of age with very few cases have been reported in pediatric age group.^{2,3,6-8} It does not have predilection for any gender.³ Exposure to radiation and chemicals like thorium, arsenic, vinyl chloride are considered for risk factors of angiosarcoma.⁹ Few authors have concluded that PSA may be result of malignant transformation of other benign splenic tumors like hemangiomas, lymphangiomas and hemangioendotheliomas.^{2,3,8,10,11}

The most common symptom in patients with PSA is abdominal pain. Other constitutional symptoms like fever, fatigue, weight loss is less frequent.^{10,12,13} Hemoperitoneum due to nontraumatic splenic rupture resulting in acute abdominal pain and distention may be a presentation in about 30% of the patients of PSA.¹⁴ Christopher et al reported a case of PSA presenting with idiopathic thrombocytopenic purpura.¹⁵ It is difficult to accurately diagnose PSA on any radiological investigation due to similar findings are seen in other splenic tumors like hemangioma, littoral cell angiomas, lymphangiomas, hemangiopericytoma.¹⁶ Ultrasound may show splenomegaly with multiple masses or single mass which may be having increased or decreased echogenicity and increased flow on doppler. There may be free fluid in the abdomen if there is rupture of spleen. CT scan usually reveals splenomegaly, diffuse infiltration of homogeneous or heterogeneous single or multiple masses with or without calcification. CT scan may also demonstrate liver, lung and bony metastasis, splenic rupture.⁷ However, FDG PET can reliably differentiate

between benign and malignant splenic masses.¹⁷ On macroscopic examination splenomegaly is always seen and diffuse involvement of spleen with replacement of entire splenic parenchyma being common feature. On microscopic examination, the tumor consists of disorganized anastomosing vascular channels lined by plump, atypical endothelial cells with large, irregular, hyperchromatic nuclei and a high mitotic rate. Naka et al in a study of 55 cases of angiosarcoma found that mitotic count and tumor size are prognostic factors.¹⁸ Due to invariable histological features, malignant vascular neoplasms of spleen may be mistaken with benign vascular tumors or malignant nonvascular tumors.^{3,6} PSA is very aggressive tumor and metastasis occurs in about (69-100) % of the cases with liver, lungs, bone and lymph nodes being commonest sites.³ Splenectomy before its rupture is the best possible treatment. Overall survival in PSA is about 4 to 14 months while 80% of the patients of rupture spleen survive not more than 6 months.¹⁹ However, Bilski et al reported a case with postoperative survival of a patient of PSA with splenic rupture for 23 months with adjuvant radiochemotherapy.²⁰ Kohutek et al also observed prolonged survival (more than 10 years) in their patient after surgery with use of postoperative radiotherapy and bisphosphonates, doxorubicin and paclitaxel based chemotherapy at three different times.²¹

CONCLUSION

PSA is very aggressive tumor which should be treated with splenectomy as early as possible. High index of suspicion and extensive workup is required to clinch the diagnosis. Splenectomy without spillage is curative.

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