

**Case Report****PRIMARY ANGIOSARCOMA OF THE SPLEEN- A RARE CASE REPORT****T. Allwyn Yabesh*, Ramya, T. Raja**

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ABSTRACT

Primary angiosarcoma of the spleen is an extremely rare and aggressive malignant neoplasm of vascular origin associated with poor prognosis. The majority of cases present with splenic rupture and hemorrhage. Here is the case report of 46 year old male who had 2 months duration of left abdominal fullness and pain. CT abdomen showed mass infiltrating spleen and liver. PETCT whole body showed features suggestive of primary splenic angiosarcoma with liver metastases. Patient underwent splenectomy and histopathology confirmed the same diagnosis. Patient was started on postoperative chemotherapy with Ifosfamide and Adriamycin.

1. Introduction

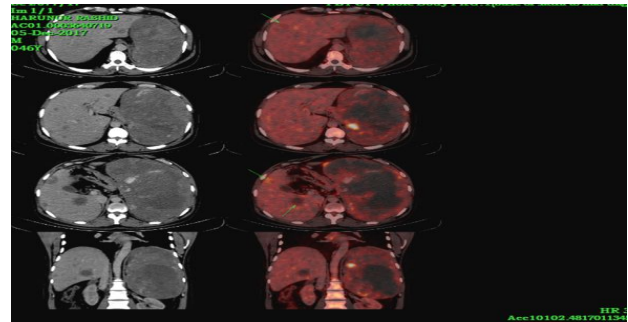
Primary angiosarcoma of the spleen is an extremely rare, highly malignant neoplasm, the pathogenesis of which is unknown, with early systemic metastatic spread and a dismal prognosis, regardless of the treatment regimen. The disease is one of the least common types of cancer, with a reported incidence of 0.14–0.25 cases per million individuals[1,2]. The majority of cases have median survival rates ranging between 4 and 18 months. Splenic angiosarcoma was first described by Langhans in 1879[3] and to date, ~300 cases have been reported worldwide. The most common sites of metastases include the liver, lungs, lymph nodes and gastrointestinal tract. Symptoms of abdominal pain and anaemia are commonly reported. Splenic rupture is a serious complication of the disease, which is frequently observed in patients, those results in mortality in a significant proportion of cases[4-6].

2. Case report

Here is a case report of 46 years old male from Bangladesh who is evaluated in hometown for the complaints of left abdominal fullness and dragging pain for 2 months. Contrast enhanced CT whole abdomen showed large heterogeneously enhancing mass lesion with blush of contrast within the lesion in the left side of abdomen which has splenic origin, compressing and displacing the adjacent structures.

Numerous centripetal enhancing lesions in liver – suggestive of splenic angio-sarcoma with hepatic metastases.

Patient came to our institution for further management. On examination patient was pale with abdominal distension involving the left hypochondrium. Spleen palpable upto right iliac fossa and non tender. Hemogram showed Hb 7.8 g/dl, Total count 13500 cells/cumm, Platelet count 139000 cells/cumm.

**Figure No. 1:** PETCT of whole body.

PETCT whole body showed peripherally hypermetabolic spleen with gross enlargement measuring 21.5 cms. The splenic parenchyma is replaced by necrosis and heterogeneously enhancing solid areas. Heterogenous FDG uptake is noted in the solid components (SUV max ranging from 4.1-6.0). Increased vascularity is noted within the mass. Liver measures 15.5 cms. Multiple well defined hypodense

lesions are seen in both lobes, largest in segment VI measures 3.5 x 3.0 cms with few of them showing heterogenous metabolic activity (SUV max 3.9). No intra hepatic biliary radicle dilatation. Portal vein, hepatic veins and IVC are normal. Imaging is in favor of splenic angiosarcoma with hepatic metastases.

He underwent splenectomy and liver nodule biopsy. Pathological examination showed external surface of the spleen is congested and focally breaching the capsule measuring 5 x 3 cm and 4 x 2 cm. Cut surface of the entire spleen is grey brown to reddish brown, firm to spongy with haemorrhagic areas. Focally there is tan brown firm area spanning an area 7 x 6 x 6 cm. Cut surface of liver tissue is grey tan, homogenous with focal grey brown haemorrhagic areas.

Sections show splenic parenchyma with extensive areas of infarction with ghost like cellular outlines of vascular spaces with haemorrhage within the lumen. The patchy viable areas show an astomosing vascular channels with focal papillary pattern lined by atypical endothelial cells exhibiting moderate pleomorphism and hyperchromasia. Also seen are cellular areas of long fascicles of spindle shaped cells with slit like spaces. Mitosis of 7-8/10 hpf is seen. Areas of necrosis are present. Immunohistochemistry stains are positive for Vimentin, CD34, CD31 and negative for Cytokeratin, HHV8. Sections show liver parenchyma with a nodule showing dilated vascular spaces with a clearance of 0.1 cm from the inked margin. Histopathology is consistent with angiosarcoma of spleen, grade 2 with Metastases to the liver.

Postoperative course of the patient was uneventful except for one episode of low grade fever.

He received pneumococcal, meningococcal and H.influenza B vaccinations in view of post splenectomy status. Patient was planned for postoperative chemotherapy and was started on Ifosfamide and Adriamycin. Patient tolerated the initial chemotherapy well and he wished to continue further cycles of chemotherapy in hometown.

3. Discussion

Primary angiosarcoma of the spleen is an uncommon and aggressive malignant neoplasm derived from the splenic vascular endothelium and elongated endothelial cells of mesenchymal origin which line the spongy network of sinusoids within the spleen. Angiosarcomas are rapid proliferating and highly infiltrating anaplastic tumors. They tend to recur locally and demonstrates wide dissemination and have an increased rate of lymph node and systemic metastases[7-11].

The etiology of primary splenic angiosarcoma remains unknown. Causes of this disease have been reported as exposure to ionizing radiation or chemotherapy for lymphoma, but no clear relationship exists[1,2,9-12]. Exposure to certain chemical agents, such as vinyl chloride, thorium dioxide and arsenic have been indicated due to their association with hepatic angiosarcomas .

The clinical manifestations of primary splenic angiosarcoma are variable, including abdominal pain, splenomegaly, anemia, fatigue, generalized weakness, fever, weight loss and even life-threatening hemorrhage resulting from rupture of the spleen. Left upper abdominal pain is the most common symptom. Constitutional symptoms common in malignancy, such as fever, fatigue and weight loss, have also been observed but are the initial symptoms <10% of the time[1,2,9-12]. The most common physical examination is splenomegaly.

Anemia is the most common laboratory abnormality, being found in 75–81% of cases, although 10–40% of patients exhibit thrombocytopenia; leukocytosis is also often noted[1,2,9,10]. One of the serious complications that is frequently observed in patients with splenic angiosarcoma is splenic rupture, leading to a fatal outcome in a significant percentage of cases, 13 to 32%.

Imaging modalities are invaluable for the differential diagnosis from other benign and malignant splenic tumors, but diagnostic accuracy is lacking. CT scans may reveal splenic enlargement with hypo- or hyper-attenuating areas . On contrast-enhanced CT scans, the tumors may show peripheral or heterogeneous contrast enhancement similar to that of hepatic cavernous hemangiomas. There is no particular pattern of calcification associated with splenic angiosarcoma, but areas of hypervascular metastases to the liver, lungs, bones, and lymphatic system are well demonstrated on CT[2,7,8] However, the clinical features and radiological appearance of the majority of cases are quite varied and non-specific, and may be easily associated with other pathological conditions, which makes early diagnosis and treatment difficult .

The usual therapy for ruptured splenic angiosarcoma is an emergency splenectomy. At present, there is no convincing evidence to suggest a clinical benefit of chemotherapy in the treatment of splenic angiosarcoma[2,12]. When considering early hematogenous micrometastasis, systemic chemotherapy following surgery may be theoretically beneficial. Although there is no standard treatment regimen, certain drugs, including cyclophosphamide, doxorubicin, epirubicin, ifosfamide, daunorubicin, vincristine, actinomycin D, steroids and Taxol, which have shown efficacy for treating angiosarcoma in other anatomical sites or other types of soft-tissue sarcomas, have been empirically proposed to be effective for primary splenic angiosarcoma.

The prognosis of patients with primary splenic angiosarcoma is very poor. Even when the primary tumor has been removed, the majority of patients succumb to systemic metastases within 1–2 years of diagnosis. Long-term survival has only previously been reported in patients who presented without metastases. Due to high risk of peritoneal or vascular dissemination, certain studies have considered splenic rupture to be the worst prognostic factor. This finding is confirmed by the fact that early splenectomy prior to rupture of the organ is accompanied by better survival rates[1].

4. Conclusion

Primary splenic angiosarcoma is an extremely rare soft tissue sarcoma. It is an aggressive soft-tissue sarcoma due to the presence of early systemic metastases and life-threatening hemorrhage from the rupture of the spleen. Pathogenesis of the disease remains unclear. The clinical and radiological diagnoses are challenging. The diagnosis of primary splenic angiosarcoma should be suspected in any patient with splenomegaly and anemia of unknown etiology. Radiation and chemotherapy have historically been unsuccessful in improving outcomes in this patient population. Further evaluation of these options will be required due to the limited experience to date. The best chance for survival follows an early diagnosis and a prompt splenectomy prior to splenic rupture.

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