

A Case of Primary Hemangiopericytoma of Liver

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Background: Hemangiopericytoma is a vascular tumor arises from pericyte of Zimmermann. Hemangiopericytoma arises from varies the location, but most commonly reported in extremities, retroperitoneum, head and neck, spine and cranium. However, intra-abdominal hemangiopericytoma is very rare. There are varies case report of intra-abdominal hemangiopericytoma including liver, spleen, pelvic cavity, and omentum; but primary hemangiopericytoma of the liver is very rare. **Case Report:** This is a case report of 49-year-old male presenting with abdominal pain for 2 months duration. No other positive history. During evaluation of the patient found to be having a mass lesion in the liver adjacent to caudate lobe, which compresses inferior vena cava. It found to be a primary hepatic hemangiopericytoma of the liver.

Key words: Hemangiopericytoma, insulin-like factors, immunohistochemistry, S-100 positive, radiotherapy

INTRODUCTION

Hemangiopericytoma is a tumor arises from the pericyte of Zimmermann. Hemangiopericytoma is a vascular tumor constitutes <1% of all vascular tumors.¹ Hepatic hemangiopericytoma can be primary or metastatic. Primary hepatic hemangiopericytoma is very rare.² Commonly affect adult at the age of fifth or sixth decades. The most common sites for hemangiopericytoma are extremities followed by retroperitoneum. Hemangiopericytoma of liver is rarely reported <1%.³

CASE REPORT

A 49-year-old male patient admitted with a complaint of abdominal pain for 2 months duration. No history of nausea, vomiting. No history of abdominal distension, jaundice, melena or bleeding per rectum. No history of fever, loss of appetite, and lose of weight. No history of previous surgery or surgical intervention. No history of prolong drug intake.

On examination, patient was conscious, oriented, and afebrile. Abdominal examinations showed tenderness in epigastric region. No sinus, scar or dilated veins. No visible peristalsis. Hernial orifices and external genitalia were normal. On auscultations, bowel sound was present.

Investigations showed complete blood count, liver function test, and renal function test were normal. Abdominal ultrasound showed well-defined solid echogenic lesion of about 5.4 cm × 3.4 cm × 3.2 cm present in the plane between the aorta and inferior vena cava [Figure 1]. No calcifications are seen. Contrast computerized tomography showed well-defined hypo dense lesion present in the caudate lobe of the liver, which enhances with contrast administration with inferior vena cava lumen narrowing of 90-95% [Figures 2 and 3]. Magnetic resonance imaging (MRI) showed lesion of 5.5 cm × 4.4 cm × 4.2 cm which showed T1-weighted low signal and T2-weighted bright signal lesion located adjacent to caudate lobe of liver, causes compression of inferior vena cava. Features are suggestive of hemangiopericytoma of the liver [Figures 4 and 5].

Preoperative diagnosis made as hemangiopericytoma and laparotomy done. Intraoperative findings were mass lesion

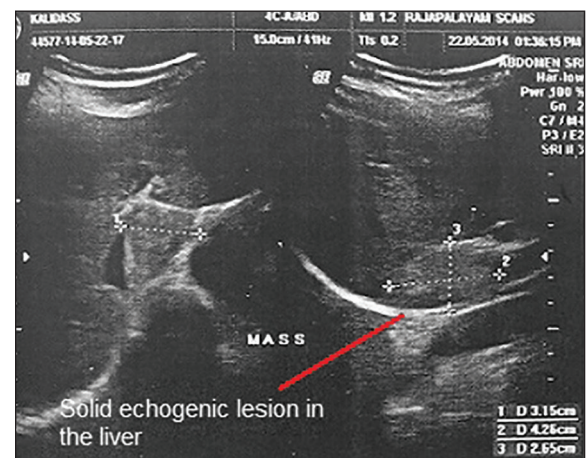


Figure 1: Ultrasonography shows solid echogenic lesion in the liver

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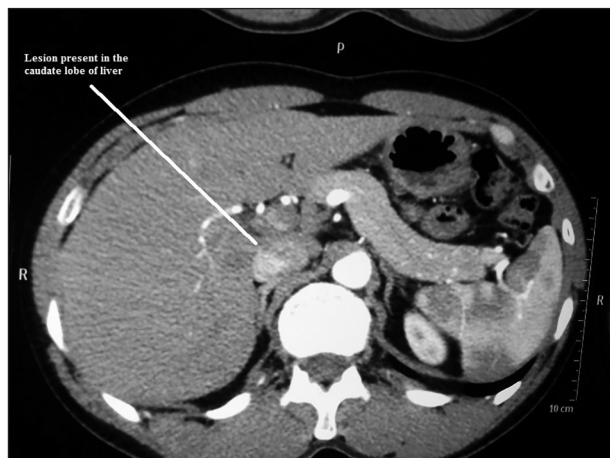


Figure 2: Computerized tomography showed well-defined hypo dense lesion present in the caudate lobe of liver

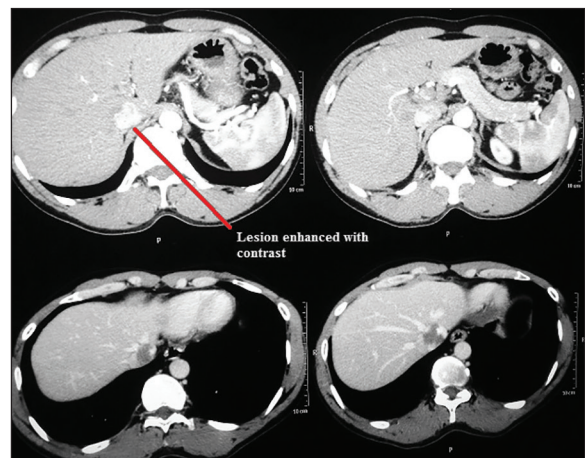


Figure 3: Computerized tomography showed well-defined hypo dense lesion present in the caudate lobe of liver, which enhances with contrast administration

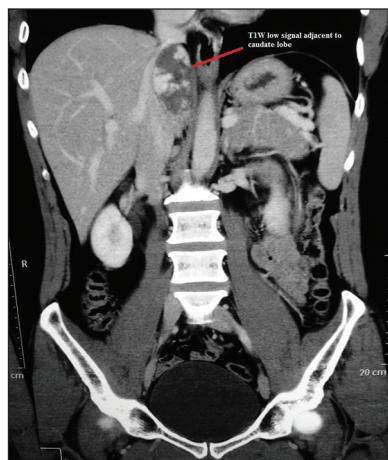


Figure 4: Magnetic resonance imaging showed lesion, which is T1-weighted low signal located adjacent to caudate lobe of liver

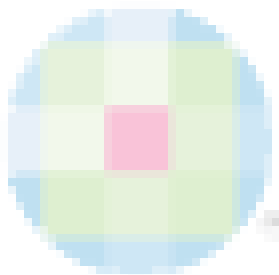


Figure 5: Magnetic resonance imaging showed lesion, which is T2-weighted bright signal lesion located adjacent to caudate lobe of liver, causes compression of inferior vena cava

arising from the caudate lobe of liver with compression to inferior vena cava [Figure 6]. No evidence of any lymph node involvement. Stomach, duodenum, and pancreas were normal. Partial hepatectomy was done. Postoperative period was uneventful. Pathology revealed a mass lesion of 6 cm × 5 cm × 4 cm, which showed features of hemangiopericytoma [Figure 7]. Resected margin was free of tumor. The tumor mass showed positive Immunohistochemistry for hemangiopericytoma.

DISCUSSION

Hemangiopericytoma may be associated with prolonged steroid intake, trauma or hypertension. It can be both benign and malignant, but malignant potential were high compared to benign.⁴ Clinical features highly variable. It can present with asymptomatic lesions to metastatic lesions. Some of the hemangiopericytoma produces hypoglycemic episodes due to

release if insulin-like factors.⁵ Some of tumor can present as acute abdomen due to rupture.⁶ The most common sites for metastasis are lung, liver, and bone. Management consists of hepatic resection with chemotherapy. Recurrent rate was very high in hemangiopericytoma.⁷

Pathological features

Hemangiopericytoma can be solitary or cystic with the well-defined pseudo capsule. Hemangiopericytoma contains numerous dilated vascular spaces with areas of hemorrhage and cystic degeneration. Immunohistochemistry is positive for vimentin, S-100, smooth muscle actin, muscle-specific actin, CD34, and factor XIIIa.^{8,9}

Computerized tomography, MRI, and angiography can be used as investigations for hepatic hemangiopericytoma.¹⁰

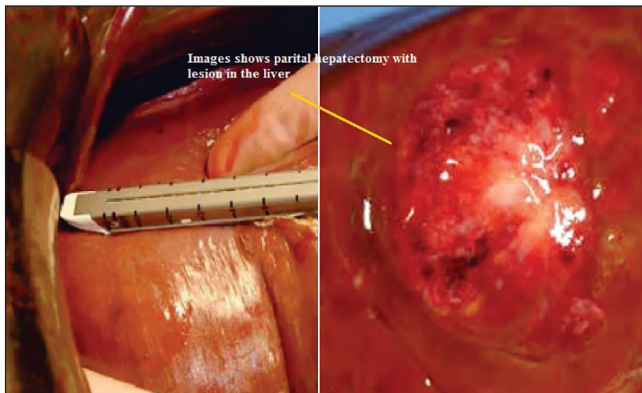


Figure 6: Intraoperative findings were mass lesion arising from caudate lobe of liver

Primary hepatic hemangiopericytoma on contrast computerized tomography shows lobulated mass with enhancing solid components, cystic areas, and speckled calcification may be suggestive of hemangiopericytoma, but not confirmative.¹¹

Treatment

Surgery is the main modality of treatment. Whenever possible tumor has been completely removed. Complete removal leads to less local recurrence and prolonged survival. Surgical modality includes segmental resection, hepatectomy or hepatic transplantation.¹²

Radiotherapy has a significant role in hemangiopericytoma. Both pre- and post-operative radiotherapy has a role in hemangiopericytoma. Preoperative radiotherapy can be given for those patients where the tumor is inaccessible for surgery, highly vascular, and large tumor. Postoperative radiotherapy can be given for those who have the possibility of local recurrence. Postoperative radiotherapy leads to decreased recurrence.¹³

Chemotherapy mostly given as palliative procedure. The most commonly used agents are actinomycin D, adriamycin, vincristine, and cyclophosphamide and methotrexate. Preoperative chemoembolization also helpful to decrease the vascularity and enhance complete excision.

Other modalities of treatment are monoclonal antibodies against vascular endothelial growth factors such as temozolomide and bevacizumab can be used.

CONCLUSION

Hemangiopericytoma is a rare vascular tumor. Hemangiopericytoma arises from the caudate lobe of the liver was less documented. It should be considered one of the differential diagnoses of the liver mass lesion, which enhances with contrast administration. Being a vascular tumor lymph

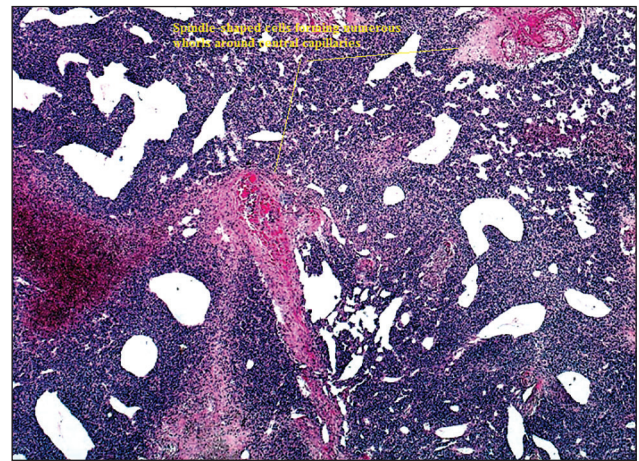


Figure 7: Pathology revealed a mass lesion of 6 cm × 5 cm × 4 cm, which showed spindle-shaped cells forming numerous whorls around central capillaries

node involvement is less. Hence, there will be good prognosis if resected margin was free of tumor.

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