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Clinical Profile and Surgical Management of Primary Hepatic Angiosarcoma: A Case Report and Review of Current Approaches

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Abstract: ***This article presents a case report of a 40 - year - old male diagnosed with primary hepatic angiosarcoma, a rare and aggressive malignancy of vascular endothelial origin. The patients atypical age of presentation and clinical symptoms, including right hypochondrial pain and a palpable lump, underscore the diagnostic challenges associated with this disease. The article discusses the challenges in differentiation from other hepatic tumors, the diagnostic role of imaging and histopathology, and the importance of immunohistochemical markers like CD31. Surgical resection remains the primary treatment option, while adjuvant therapies efficacy is limited. The case highlights the need for further research to improve treatment strategies for this highly lethal malignancy.***

Keywords: Primary hepatic angiosarcoma, surgical resection, diagnostic challenges, immunohistochemistry, adjuvanttherapy

1. Introduction

Hepatic angiosarcoma is a very rare primary liver malignancy disease, accounting for only 2% ofprimary liver malignancies. It arises from stromal vascular endothelial cells. Though the etiology of primary hepatic angiosarcoma remains unclear, chronic exposure to chemicals, such as thorium dioxide, arsenic, vinyl chloride, and radium, has been associated with it [2, 3]. Patient usually presents with pain or lump in right hypochondrium or non specific symptoms like loss of appetite and weight loss. Till date, the therapeutic guidelines for liver angiosarcoma has not been set up; partial liver resection to remove tumor radically still remains to be the cornerstone of treatment followed by adjuvant chemotherapy. Primary hepatic angiosarcoma has very poor prognosis which is attributable to its rapid progress, high recurrence rate, and resistance to traditional chemotherapy (4). Here we present a case report of 40 years old male diagnosed as primary hepatic angiosarcoma who underwent partial liver resection.

2. Case Report

40 years old male presented with chief complaint of pain in right hypochondrium, dull aching, moderate to severe, relieved on medication since 1 month. Patient also felt lump in epigastrium since 1 month. On examination there was tenderness in right hypochondrium with hepatomegaly. CEMRI abdomen was done and findings were suggestive of enlarged liver span of 21 cm with the presence of two large thick walled multiloculated cystic lesions in left lobe of liver with internal septations and fluid fluid level within it and post contrast peripheral heterogenous enhancement. Left hepatectomy was done and operative findings were suggestive of large 20 X 15cm bilobed solid cystic lesion in left lobe of liver. Hemorrhagic fluid was aspirated from the lesion. Histopathological examination revealed 2 well circumscribed multiloculated solid cystic lesions with multiple areas of hemorrhage and necrosis. Dilated cavernous sinuses were lined by tumor cells reaching till glisons capsule but not infiltrating it. Resection margins were free of tumor cells. On immunohistochemistry, tumor cells were positive for CD31 while negative for CD34, S100, SMA and MDM2. MIB - 1 was 40% in highest proliferating areas. After surgical resection, patient recovered very well and was discharged two weeks later. Post operative contrast enhanced CT scan abdomen showed no residual or recurrent tumor. Patientrefused for adjuvant chemotherapy and is on follow up with good performance status.

3. Discussion

Primary hepatic angiosarcoma is an aggressive malignant tumor arising from vascular endothelium with a low incidence rate and a dismal prognosis. It is usually seen in males and most common age of presentation is more than 60 years. However, in our case the age of presentation was 40 years. The presenting symptoms are usually non specific with abdominal pain, discomfort, loss of appetite, weight loss being more common. In our case the patient presented with pain and lump in right hypochondrium since 1 month. Tumor markers are usually within normal limits. Imaging helps in establishing diagnosis, but it is confirmed by histopathological examination only. Due to the hypervascular characteristic of liver angiosarcoma, it is difficult to differentiate liver angiosarcoma from other vascular tumors in liver, such as hepatoma or adenoma, radiologically. On histopathology, the resected liver shows tumor composed of anastomosing vascular channels infiltrating surrounding hepatic tissue in a destructive fashion. Immunohistochemically, the tumor is positive for CD31, CD34, and Vimentin which is a mesenchymal marker. However in our case only CD 31 was positive. At present, the cornerstone of treatment for liver angiosarcoma remains surgical resection. Adjuvant chemotherapy or radiotherapy fails to reach a survival benefit [5]. In our case, young male patient with primary hepatic angiosarcoma

underwent left hepatectomy and subsequently kept on follow up in view of no residual tumor on post operative CT scan and consent for adjuvant chemotherapy not being given by patient. At present, the patient is asymptomatic with good performance status with 6 months of follow up. Despite of treatment, only 3% of patients are reported to live longer than 2 years [6]. The survival of patients with liver angiosarcoma is very poor due to its non specific symptoms at presentation, high recurrence rate and high probability of spontaneous rupture and tumor spillage.

4. Conclusion

Primary hepatic angiosarcoma is an aggressive malignancy with dismal prognosis. Till date surgery remains the mainstay of treatment. Due to Low incidence rate, no standard guidelines have been established on adjuvant therapy yet and hence it seeks further research in treatment options.

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