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CASE REPORT

HEPATIC ANGIOSARCOMA: REPORT OF A CASE WITHOUT A HISTORY OF EXPOSURE TO ENVIRONMENTAL RISK FACTORS

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Abstract

A 30 year-old male patient was referred with RUQ abdominal pain of 10 months duration. With the impression of acute abdomen, the patient underwent surgery. The pathologist proposed hydatid cyst as the probable diagnosis. Because of persisting abdominal pain the patient was reevaluated. CT scan showed a mass lesion, 22cm in diameter, in the right hepatic lobe and thus right hepatic lobectomy and cholecystectomy were performed. Histologic findings were consistent with angiosarcoma. Increased familiarity with this cancer will facilitate correct diagnosis and will help to differentiate it from the more prevalent hepatic mass lesions found in Iran.

Keywords • Angiosarcoma • liver • cancer

Introduction

Primary hepatic angiosarcoma is a rare mesenchymal tumor of the liver that is usually related to history of exposure to environmental carcinogens. Recent series show that this tumor is not uncommon in young individuals lacking associated risk factors.[1] Here, we present a case of primary liver angiosarcoma in a young patient without obvious risk factors, with emphasis on the fact that one should consider this tumor in the differential diagnosis of liver masses even in the absence of all risk factors.

Case report

A 30-year-old male construction worker presented with RUQ colicky abdominal pain which radiated to the right shoulder, nausea, vomiting, anorexia and progressive weight loss which had begun 10 months prior to his admission to our center. Ultrasound studies revealed a huge abdominal mass with mixed echogenicity in the right hepatic lobe. The patient underwent laparotomy when he developed signs of acute abdomen. Coffee-ground fluid with blood clots and necrotic material were discharged from this mass during the operation and pathology showed uncertain evidence of hydatid cyst (cyst wall with surrounding inflamed hyalinized connective tissue and no scolex was present). The drainage tube was placed and maintained for 45 days. After one month, abdominal pain was still present and physical examination revealed splenomegaly and tender hepatomegaly. CT scan showed a mass lesion of 22 cm in diameter, in the anterio-superior segment of the right hepatic lobe, which had displaced the right hemidiaphragm and right kidney. The mixed density of the lesion indicated probable complicated hydatid cyst. Right hepatic lobectomy and cholecystectomy were performed.

Microscopic examination revealed a vascular tumor with undifferentiated plump spindle shaped anaplastic endothelial cells producing some vascular channels. These cells were spindle-shaped or pleomorphic in appearance and they had hyperchromatic nuclei. Multiple nucleoli and meiotic activity were seen. The postoperative course was uneventful for two months and the patient was referred to an oncologist.

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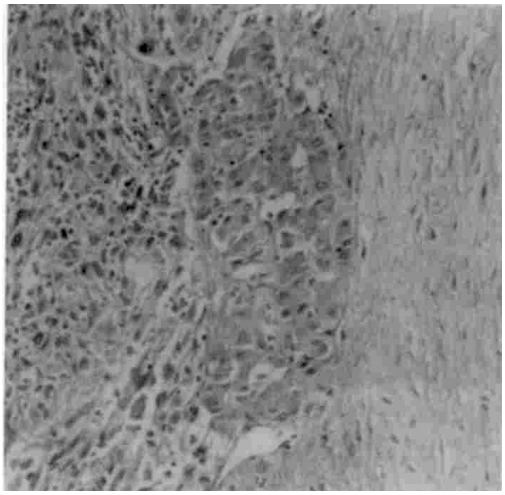

Figure 1. Low power photomicrograph showing liver tissue adjacent to neoplastic cells.

Discussion

Angiosarcoma (also referred to as malignant hemangioendothelioma, hemangiosarcoma or kupffer cell sarcoma) is a rare liver tumor with a rapidly fatal course and it is the most common malignant mesenchymal tumor affecting the liver.[2] Patients range in age from 24 to 93 years with the peak incidence in the 6th and 7th decades; 85% of the patients are male.[2] The most common initial symptom is abdominal pain and other common symptoms include abdominal enlargement, nausea, anorexia, occasional vomiting and jaundice.[3] The most common physical sign is hepatomegaly with ascites. Splenomegaly is seen less often.[4] The reported patient had bouts of abdominal pain mimicking acute abdomen. Common complications include portal vein thrombosis, Budd-Chiari syndrome, arteriovenous or arterioportal shunts[1] and rupture which is reported in 15% of cases. Also five cases of idiopathic portal hypertension have been reported in association with hepatic angiosarcoma.[5] In this case, 500 mL of blood was found in the pelvic cavity which may indicate partial rupture of the tumor.[6]

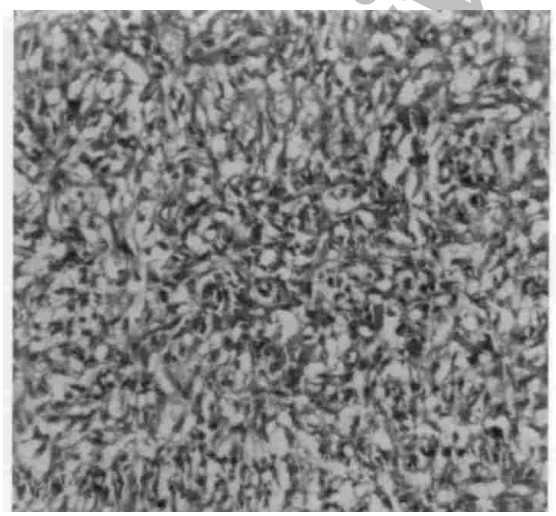

Figure 2. High power photomicrograph revealing a highly vascular tumor with anaplastic cells and numerous tortuous vascular channels.

Angiosarcoma has become a subject of interest because of its close relationship with environmental carcinogens, such as thorium dioxide (thorotrast), vinyle chloride monomer and different arsenical solutions.[2] Hemochromatosis is also mentioned as a potential predisposing factor. This patient denied history of contact with any of the aforementioned carcinogens and his medical records revealed no potential risk factor. Combined use of angiography and dual-phase helical CT provides a better identification of the tumor and its complications and should be used when the clinical suspicion is high.[1] Angiosarcoma of the liver is a multifocal tumor with a variety of findings on multiphasic contrast-enhanced helical CT.[7] Due to vascularity of the tumor, percutaneous biopsy is hazardous.[1]

Being an aggressive neoplasm, most patients susually die within the first 6 months of diagnosis and distant metastases develop in 50% of patients.[8] Surgical resection is usually unsuccessful because most patients present in the advanced stage of the disease. Some long-term survivors have been reported after a partial hepatectomy but a few patients survive more than 1 to 3 years after its complete resection. Attempts to treat hepatic angiosarcoma with radiation therapy and/or chemotherapy is disappointing.[9]

The points worth highlighting in this report are the diagnosis of liver angiosarcoma in a 30-year-old male without any risk factors and detecting the probable pitfalls in the diagnosis and management of such patients. Considering the high prevalence of hydatid disease in Iran, one should consider the diagnosis of angiosarcoma in the differential

diagnosis of atypical cases of this disease.

References

1 Rademarker J, Widjaja A, Galanski M. Hepatic hemangiosarcoma imaging findings and differential diagnosis. *Eur Radiol*. 2000; **10:** 129-33.

2 Devita VT, Hellman S, Rosenbery SA. *Cancer: Principles and Practice of Oncology*. Vol 1. 5th ed. Philadelphia: Lippincott-Raven; 1997: 108.

3 Zakim D, Boyer TD. *Hepatology: A Textbook of Liver Disease*. 2nd ed. Philadelphia: Saunders; 1990: 1224-8.

4 La Dou J. *Occupational Medicine*. Norwalk: Appleton and Lang; 1990: 200-1.

5 Duenus C, Perez-Alvarez JC, Busteros JI, et al. Idiopathic portal hypertension and angiosarcoma associated with arsenical salt therapy. *J Clin Gastroenterol*. 1998; **26:** 303-5.

6 Tsai MH, Chien RN, Hsieh SY, Hung CF, Chen TC, Sheen IS. Primary hepatic angiosarcoma: report of a case involving environmental arsenic exposure. *Changging Yi Xue Za Zhi*. 1998; **21:** 469-74.

7 Peterson MS, Baron RL, Rankin SC. Hepatic angiosarcoma: findings on multiphasic contracts-enhanced helical CT do not mimic hepatic hemangioma. *AJR Am Roentgenol*. 2000; **175:** 167-70.

8 Damjanor I, Linder J. *Anderson's Pathology*. 10th ed. St Louis: Mosby; 1996: 1838-9.

9 Bell RH, Kikkers LF, Mulholland MW. *Digestive Tract Surgery: A Text and Atlas*. Philadelphia: Lippincott-Raven; 1996: 642-93.