An unusual presentation of hemangiomatosis of the liver

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ABSTRACT

Diffuse hemangiomatosis of the liver is a rare condition of infancy characterized by extensive hemangiomas in the liver. It is extremely rare in adults. The imaging findings are similar to hemangioma showing centripetal enhancement with stasis of contrast. We present a case of hemangiomatosis of the liver and spleen in a 52 year old male patient. Following splenectomy there was enhanced growth of the hepatic hemangiomatosis within a period of four months and died soon after due to liver failure. We could not find a similar rapid progression of hepatic hemangiomatosis in our literature search.

Keywords: CT, Liver neoplasm, MRI, hemangioma.

Keymessages: Hepatic hemangiomatosis is a rare benign condition, that can occur with exposure to drugs like oestrogen. Sometimes it can occur without exposure to drugs and can be rapidly progressive and lethal.

INTRODUCTION

Diffuse hepatic hemangiomatosis is an extremely uncommon benign condition in which the normal liver parenchyma is replaced by tissue with histologic similarity to cavernous hemangiomas.[1] It is an angiogenesis dependent condition with periods of rapid growth (activation) followed by regression. It may be associated with usage of oral contraceptive drugs and metoclopramide.[2]

Case history

A 52 year old male was admitted to our hospital with complaints of pain in the left hypochondrium for three weeks. There was no history of intake of any drugs. Clinical examination of the abdomen revealed a massive splenomegaly. Baseline investigations showed normochromic normocytic anemia, thrombocytopenia and eosinophilia. In view of anemia, a bone marrow biopsy was done which revealed features suggestive of reactive bone marrow with eosinophilia. Sonography of the abdomen showed splenomegaly with multiple hyperechoic nodules in the spleen. Contrast Enhanced computed tomography (CT) of the abdomen demonstrated multiple enhancing nodular lesions in spleen (Figure 1). No demonstrable focal lesions were seen in the liver. In view of the multiple space occupying lesions in the spleen, laparotomy and splenectomy was performed to rule out malignancy. Histopathological examination showed features of splenic hemangiomatosis (Figure 2A). Postoperative period was uneventful and the patient was discharged.

Four months later, the patient got admitted with complaints of abdominal distension. Physical examination revealed hepatomegaly. Laboratory investigations revealed deteriorated liver function with total bilirubin of 3.5 mg/dl (Direct-2.7 mg/dl), total proteins of 7.9 g/dl (albumin-2.7 g/dl, globulin 5.2 g/dl), and hemoglobin of 9.6 g/dl. Sonography of the abdomen showed ascitis and gross hepatomegaly with multiple hyperechoic nodules (Figure 3). Contrast Enhanced CT of the abdomen was
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the patient died after 6 months due to liver failure as he was waiting for an appropriate liver donor.

DIFFUSION

Diffuse hepatic hemangiomatosis is different from multiple hepatic hemangiomas as the former is characterized by ill-defined margins whereas the latter has smooth, well-defined margins. However, the two conditions often co-exist. Although the etiology is not completely understood, it may be associated with usage of drugs like oral
contraceptives.[2] It is slightly more common in women than in men. The right lobe of the liver is more commonly involved than the left lobe, but in most cases the entire liver is involved. The patients present with a spectrum of symptoms like abdominal pain, abdominal distension, vomiting, weight loss, fever, jaundice and dyspnoea.[3–5] Diffuse hepatic hemangiomatosis can be associated with Rendu-Osler-Weber disease (Skeletal hemangiomatosis) and systemic hemangiomatosis.[1]

Sonography shows solitary or multiple hyperechoic areas with homogenous internal echoes. On non enhanced CT, it appears as a hypoattenuating lesion with ill-defined margins.[3] Some of them may have areas of hemorrhage within the tumor. On contrast administration, it shows centripetal enhancement with stasis of contrast.[3] On MR imaging the characteristic finding in hepatic hemangiomatosis is T2 hyperintensity with alternating areas of normal hepatic parenchyma.[3] The lesion appears hypointense on T1 weighted images as compared to the spleen. Arteriography shows multiple dilated vessels and pooling of contrast within the lesion.[2]

Histological examination reveals thin walled cavernous vessels lined by cuboidal endothelial cells with no evidence of cytologic atypia or mitosis.[3] Frank serge et al.[2] has described a case where it took more than two years for the disease to progress to right lobe of liver following left hepatectomy. Ozakyol et al.[3] have described development of hemangiomatosis in two women after two years following estrogen administration. However following withdrawal of estrogen there was no further growth of these lesions. In our patient, hemangiomatosis rapidly progressed in four months following splenectomy. Complications of diffuse hemangiomatosis include spontaneous rupture, thrombocytopenia, spontaneous haemorrhage, consumption coagulopathy (Kasabach-Merritt Syndrome), portal hypertension, arterio-venous shunting and cardiac failure.

The treatment of adult hemangiomatosis is not well defined because of the rarity of the disease. Corticosteroid, interferon, radiotherapy, lobectomy and hepatic transplantation can be tried to treat diffuse hepatic hemangiomatosis.[6–8] Ozakyol et al.[3] treated their two women patients by withdrawing the offending agent, namely estrogen. Eriko Maeda et al.[3] treated their two patients by liver transplant and right lobectomy. Emily et al.[8] have proposed the following treatment in infantile hemangiomatosis. They suggested that asymptomatic patients be followed up with sonography. Infants with haemodynamically significant shunting to be treated with corticosteroids and if they don’t respond, to be treated with embolisation although they don’t affect the underlying hemangiomatosis. Resection of a lobe is reserved for those patients who don not respond to the above methods. Diffuse hemangiomatosis with liver failure are to be treated by liver transplantation. In cases of splenic hemangiomatosis, splenectomy has to be done due to the increased risk of splenic rupture.[7]

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REFERENCES