

Haemangioma-thrombocytopenia syndrome – A case report

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Summary

A 29 year old Chinese female who presented with spontaneous purpura, was found to have gross hepatomegaly and thrombocytopenia. The thrombocytopenia responded to steroid therapy but relapsed when the dose of steroid was tapered down. Subsequent investigations revealed that the hepatomegaly was due to a large haemangioma of her liver. For symptomatic hepatic haemangioma, surgical excision is the treatment of choice; this was refused by the patient.

Key words: Hepatic haemangioma, purpura, thrombocytopenia.

Introduction

Large, symptomatic hepatic haemangiomata are uncommon and pose as diagnostic and management challenges to the clinician. The following is a case of the very rare haemangioma-thrombocytopenia syndrome.

Case report

CCM was a 29 year old Chinese lady who started to have spontaneous bruises on her legs in September 1985. She was seen by a physician in private practice who found her to have an enlarged liver which was palpable 10 cm below the right costal margin and a low platelet count of $50 \times 10^9/l$. A bone marrow examination showed increased number of megakaryocytes and the patient was treated with prednisolone 60 mg a day. The thrombocytopenia gradually improved and after two months of treatment with high dose steroids the platelet count increased to $150 \times 10^9/l$. By then she did not have anymore bleeding tendency. The dose of prednisolone was tailed down and this was followed by a drop in the platelet count in February 1986.

Since September 1985 she had noticed progressive upper abdominal distension with epigastric discomfort. There was no loss of appetite nor weight loss. There was no history of previous liver disease. She was unmarried and had never been on contraceptives.

In May 1986 she was referred to University Hospital, Kuala Lumpur, for investigation of her hepatomegaly and thrombocytopenia. Examination revealed that she was anicteric and there

was no pallor. There was no cutaneous haemangioma. The lower edge of her liver was 12 cm below the right costal margin. It was firm, nodular and non-tender. No bruit was heard on auscultation. Her spleen was not palpable and there was no ascites. The rest of the examination was normal. The provisional diagnosis was hepatocellular carcinoma.

Her haemoglobin was 11.6 g/dl and platelet counts ranged from $39 \times 10^9/l$ to $53 \times 10^9/l$. The peripheral blood picture was normal. The serum albumin and bilirubin were normal. The alkaline phosphatase was 166 IU/l (normal range 34–135 IU/l), aspartate transaminase was 55 IU/l (normal range 7–40 IU/l) and alanine transaminase was 69 IU/l (normal range 4–54 IU/l). The prothrombin time ratio was 1.1 and the partial thromboplastin time was not prolonged. HBsAg and alpha-fetoprotein were not detected in her serum. Bone marrow examination showed increased number of megakaryocytes. Ultrasound examination showed an echogenic mass involving the right lobe of the liver. CT scan revealed a mass lesion occupying almost the whole of the right lobe of the liver which enhanced with intravenous contrast from the peripheries inwards. Hepatic arteriogram showed that the arterial phase was normal except that the left hepatic artery did not fill up. There was marked pooling of contrast in the capillary bed of the right lobe of the liver. Delayed films did not show opacification of any hepatic veins (Figures 1 and 2). The radiological diagnosis was haemangioma of the right lobe of the liver.

The patient refused surgical treatment and subsequently was discharged.

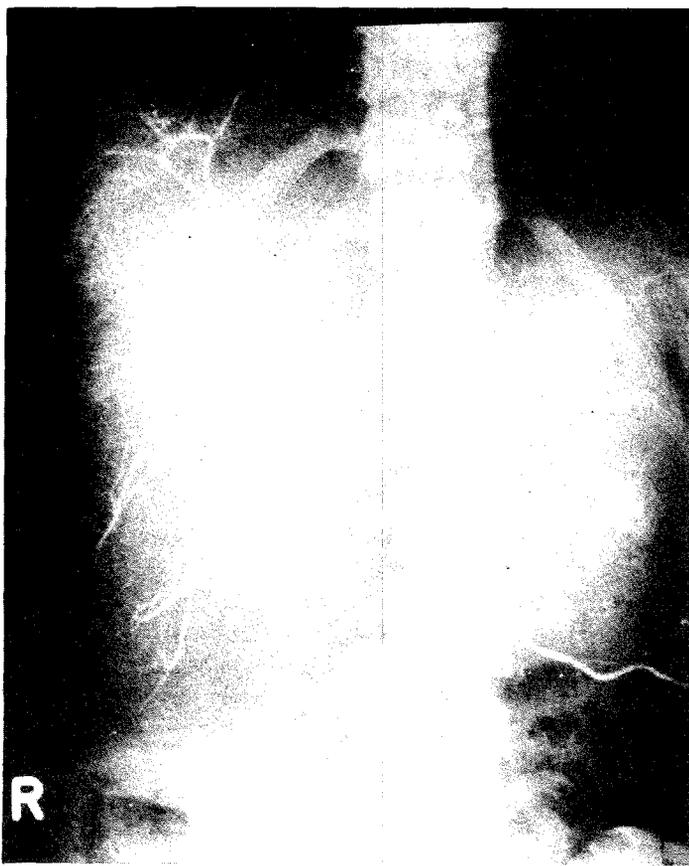


Fig. 1 Arterial phase of hepatic arteriogram.

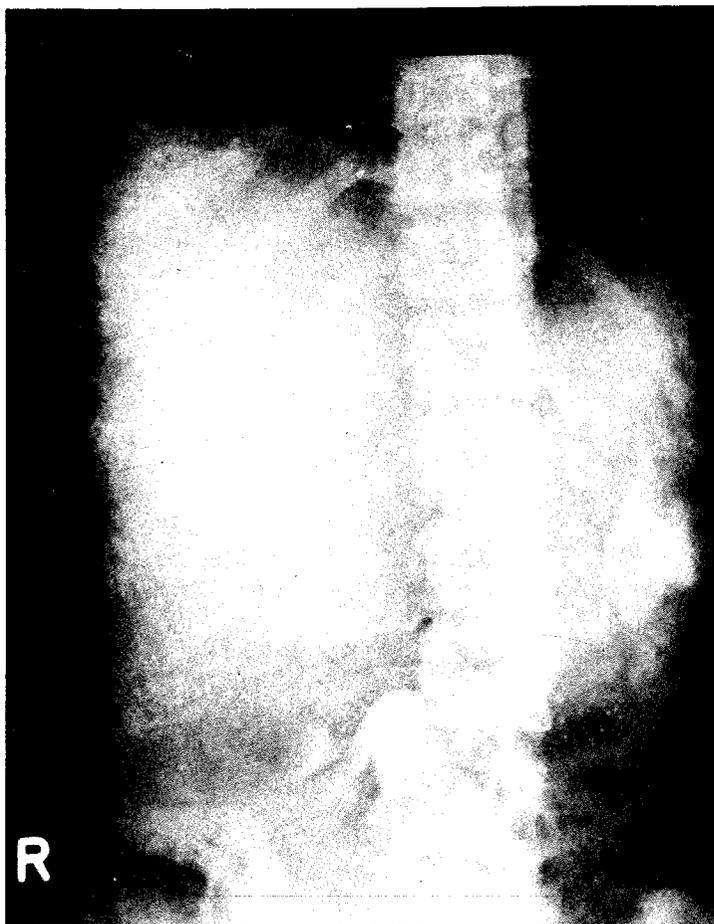


Fig. 2 Marked pooling of contrast in capillary bed of right lobe of liver.

Discussion

Hepatic haemangiomas are rare. In 2,400 autopsies reviewed by Oschsner the incidence of hepatic haemangioma was found to be 2%.¹ It is four to five times more common in females. The most common age of clinical presentation is the 3rd and 4th decades of life.

Most authorities agree that hepatic haemangiomas are hamartomatous malformations.¹ In general, these tumours are soft and cystic but the consistency may vary with the amount of thrombosis and subsequent fibrosis that may occur. Microscopically, there are large irregular spaces filled with blood, lined by a single layer of vascular endothelium and separated by connective tissue septae. Multiple haemangiomas occur in 10% of cases. Solitary lesion often affects the right lobe. They may be associated with cutaneous haemangiomas.

Only 13.5% of cases reviewed by Ishak² had clinical symptoms of which upper abdominal fullness and pressure symptoms from hepatomegaly are the commonest. The hepatomegaly is usually mistaken for malignant tumours and may be tender. A bruit is rarely detected. A high cardiac output failure state due to arteriovenous shunting, commonly seen in haemangioma presenting in infancy is rarely seen in the adult patient.

Findings of selective hepatic arteriography are characteristic. Cavernous haemangioma shows large feeding vessels which are displaced and crowded together at the edges. There are large varix-like spaces which are rapidly filled with contrast material and remain densely opacified throughout the entire angiographic examination. Needle biopsy is contraindicated because it can lead to fatal haemorrhage.

Kasabach and Merritt first described the syndrome of haemangioma with thrombocytopenia and purpura in a two month old infant with cutaneous haemangioma in 1940. Approximately 100 cases have been reported subsequently. Most of the haemangiomas were localised to the extremities. The syndrome is most commonly seen in infants but can occur in adults as well. It is believed that the thrombocytopenia is secondary to a consumption coagulopathy initiated within the haemangioma.³ Our patient did not have laboratory evidence of disseminated intravascular coagulation.

For symptomatic hepatic haemangioma surgical excision is the treatment of choice if the lesion is confined to one lobe of the liver. However, when both lobes of the liver are extensively involved radiotherapy or hepatic artery ligation are alternative modes of therapy. Steroids have been used with some success in infants with hepatic haemangiomas but the effectiveness of steroids in adults is not known. It is interesting that the thrombocytopenia in our patient responded to high dose prednisolone.

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