MASSIVE HEPATIC HAEMANGIOMATOSIS

C.H. LIM
L. TAN

SYNOPSIS

Massive hepatic haemangiomatosis are relatively rare benign vascular tumours and usually present in infancy with congestive cardiac failure or as an unexplained abdominal mass. Its occurrence in adults is uncommon as the lesions tend to involute. This is the first documented case of massive hepatic haemangiomatosis presenting with congestive cardiac failure in an adult. The haemodynamic and angiographic findings are described and discussed.

Haemangiomas of the liver first described by Virchow and Rokitansky (1863) are benign vascular tumours, usually asymptomatic and discovered incidentally at operation or autopsy. However, the massive hepatic haemangiomas usually present in infancy as unexplained congestive cardiac failure or as an abdominal mass (Dehner, 1971). Heart failure is due to an excessively high cardiac output resulting from massive intrahepatic arteriovenous shunting. The purpose of this paper is to report a case of massive hepatic haemangiomatosis in a 33 year old adult Chinese female presenting with congestive cardiac failure during pregnancy. As far as we are aware, this is the first reported case of massive haemangiomatosis presenting with cardiac failure in an adult.

CASE HISTORY

E.S.K., a 33 year old Chinese housewife developed congestive cardiac failure during the last trimester of her third pregnancy. She had an uneventful normal delivery and was referred for assessment of her cardiac status prior to tubal ligation. Her two previous pregnancies in 1972 and 1975 were normal and uncomplicated.

Physical examination showed that the blood pressure was 140/70, pulses were collapsing. The apex beat was dis-

Department of Medicine (II),
University of Singapore,
Sepoy Lines,
Singapore 3.

Lim Chin Hock, AM. MBBS, M.Med. (Int. Med.), FRACP

Lenny Tan, AM, MBBS, DMRD, FRCR.

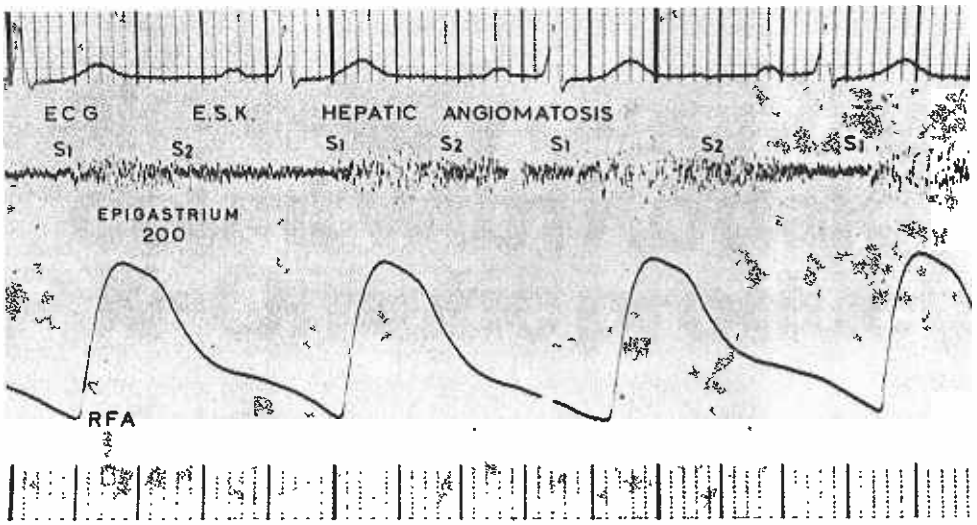

Fig. 1. A phonocardiographic record of the loud and continuous bruit heard over the epigastrium.

placed laterally to the sixth left intercostal space, anterior axillary line. Jugular venous pressure was not raised. A thrill was present over the epigastrium. The heard sounds were normal, no gallops were heard. An ejection systolic murmur was heard over the pulmonary area. A loud continuous murmur was heard over the epigastrium (Fig. 1).

The liver was enlarged to 5 cm, and the spleen to 1½ cm. There were no cutaneous lesions.

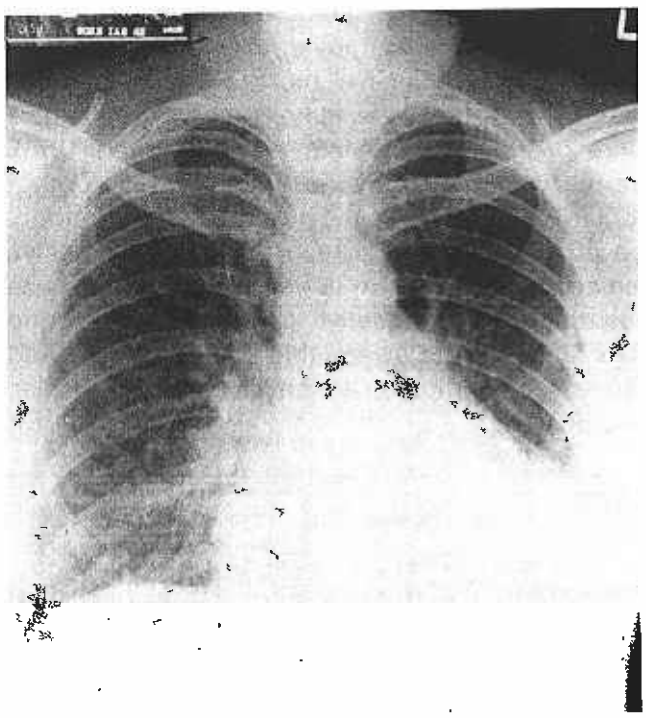

Fig. 2. Chest X-ray of the patient shows cardiomegaly, upper lobe diversion, pulmonary phethora and 'plump' pulmonary arteries.

Investigations

Hb 13.9 gm per cent, Tw 6,000/ul, platelets 230,000/ul. ESR 12 mm/1st hour. Urine analysis was normal. Liver function tests—SGPT 48 IU/L, alkaline phosphatase 9 units, total protein 4.4 gm/dl, serum albumin 2.4 gm/dl, serum bilirubin 0.8 mg per cent. ECG was within normal limits. Chest X-ray showed cardiomegaly, pulmonary plethora and evidence of upper lobe diversion (Fig. 2).

Cardiac catheterisation was done on 19.12.75 (Table I) and this revealed the presence of mild

TABLE I: Haemodynamic Findings

Site	Pressure in mmHg		Blood Samples
	Phasic	Mean	Saturation %
PCP	a = 20, X = 18 v = 24, y = 12	18	90
MPA	50/28	34	84.5
HRV			91
MRV	50/17		83
LRV			84
SVC			75
HRA			79
MRA	a = 17, x = 14 v = 15. y = 10	12	81.5
LRA			86
Abdominal aorta	130/70	92	90
LV	130/26		90

Cardiac output = 12.82 L/min
Cardiac index = 8.78 L/min/m^2

pulmonary arterial hypertension. The low systemic saturation could be due to the large veno-arterial shunting. The cardiac output was 12.82 L/min and the cardiac index was 8.78 L/min/m^2 (dye-curve method).

An abdnominal aortogram was done and this was followed by selective injections into the coeliac axis, splenic artery and intercostal arteries. The arteriograms (Figs. 3 & 4) revealed large, dilated and tortuous coeliac, hepatic and splenic arteries. The hepatic artery arose close to the origin of the coeliac axis. There was generalised mottled collection of contrast in both the right and left lobes. There was early opacification

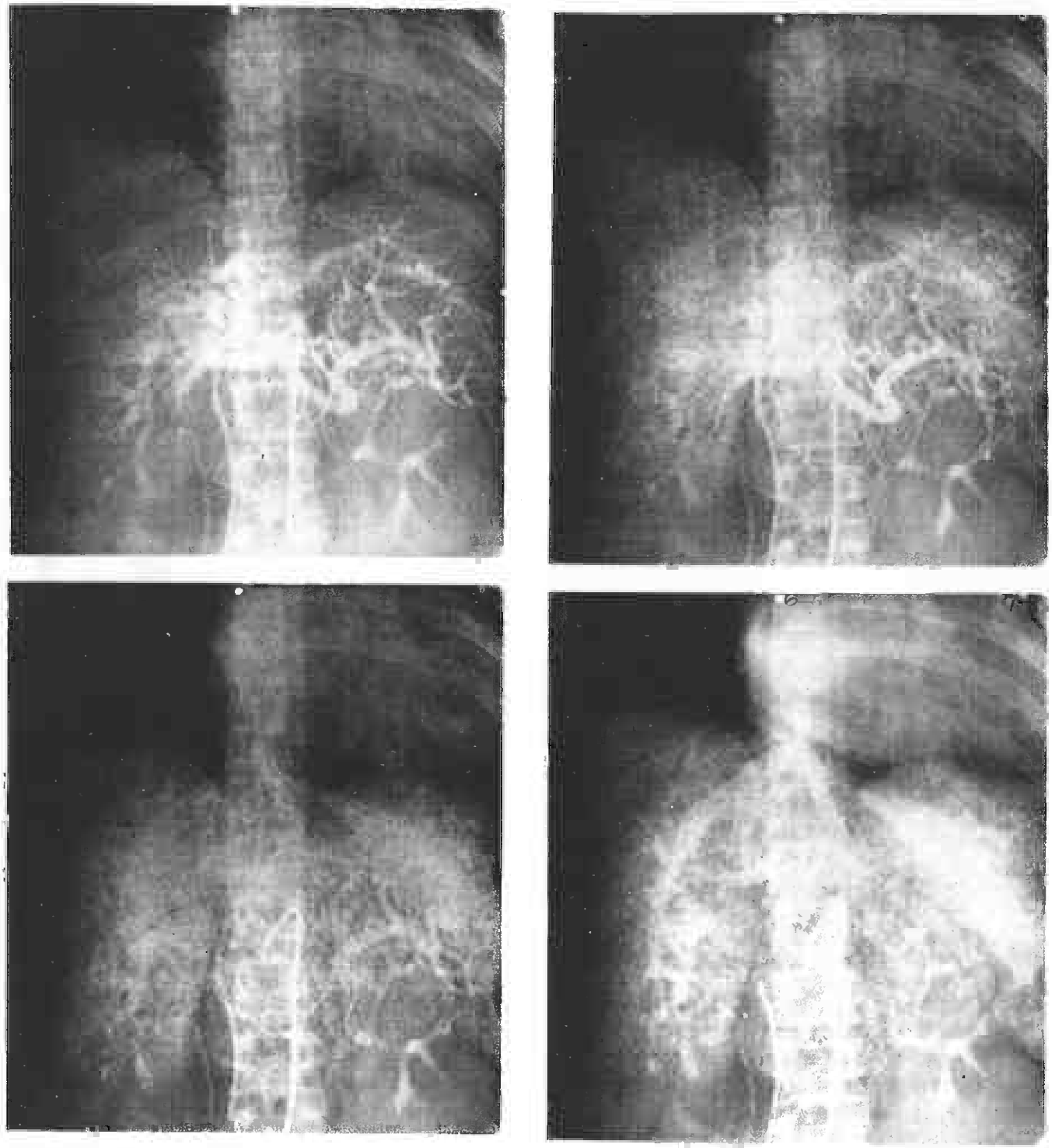

Fig. 3a-3d. This series of films show the catheter tip in the coeliac axis. The hepatic and splenic arteries are dilated and tortuous. There is a 'mottled' and blotchy appearance due to the presence of contrast in the multiple hepatic haemangiomas. Early opacification of the hepatic veins, inferior vena cava and right atrium is seen in Fig. 3c and 3d. Both kidneys are markedly enlarged.

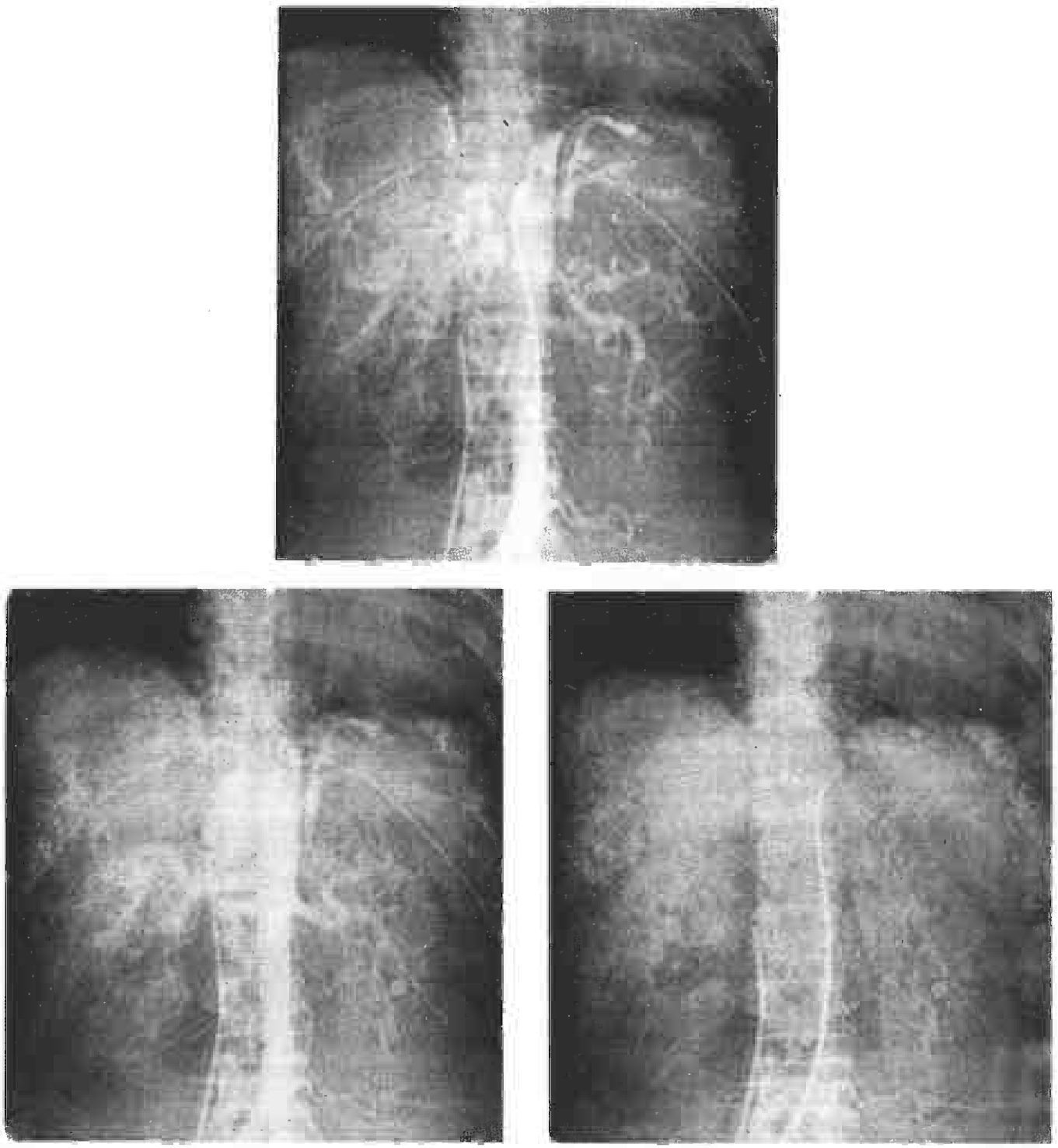

Fig. 4a-4c. The general arteriogram demonstrates the dilated and tortuous hepatic and splenic arteries. The extent of the hepatic haemangiomatosis is well seen in these films.

of the hepatic veins, and contrast could be seen draining into the inferior vena cava and the right atrium (Figs. 3a-3d). Both kidneys were large. A selective splenic arteriogram revealed the absence of haemangiomas in the spleen. There were feeding vessels from the left gastric and intercostal arteries.

Progress

Her cardiac failure improved with digoxin and diuretics. She is presently asymptomatic and her effort tolerance is normal.

DISCUSSION

Haemangiomas of the liver have been described

mainly in infants (Slovis, 1975; De Lorimer, 1967; Miller, 1972). Massive hepatic haemangiomas which produce symptoms, too are uncommon. Infants with these lesions usually present with one or more of the following manifestations—an abdominal mass, high cardiac output state or congestive heart failure. In addition, they may have cutaneous haemangiomas, cardiac murmurs, or an abdominal bruit. Isotopic scanning of the liver can be used as a screening procedure to detect this lesion. Arteriography is however the most definite diagnostic test. The giant haemangiomas are angiographically different from the cavernous type of haemangiomas. In the former, the hepatic artery is markedly dilated supplying large vascular channels throughout the tumour and there is rapid venous filling of the hepatic veins. In cavernous haemangiomas, the hepatic artery and its branches are usually not dilated and tortuous. The dilated vessels retain the contrast for a long time and well into the venous phase (Laws, 1975). In 25 cases with hepatic haemangiomas and cardiac failure, the mortality rate of untreated cases was 88 per cent (De Lorimer, 1967).

In infants, hepato-blastomas have to be excluded. Slovis (1975) suggested that the hepatic lesion is a benign vascular tumour if the patient presents with congestive cardiac failure, and if there is angiographic evidence of early drainage into the hepatic veins.

The haemangiomas which are benign vascular tumours usually regress from infancy (Miller, 1972; Slovis, 1975). Involution of these tumours occur before six months of age in 16 per cent, while 65 per cent involute between six to twelve months. Because of this tendency to spontaneous involution, supportive therapy with digitalis, diuretics, steroids (Goldberg, 1969) and radiation therapy have been described (Slovis, 1975). However, surgical treatment i.e. excision of localised lesions (Matolo, 1963), hepatic artery ligation (De Lorimer, 1967) have been described. However, the latter form of treatment is not effective as collateral flow may restore hepatic artery flow to previous pre-operative flow values. Surgery is usually reserved for those cases who have intractable cardiac failure, severe thrombocytopenia due to platelet sequestration, or rupture of the liver (Slovis, 1975).

As far as we are aware, our patient is the only documented case of hepatic haemangiomatosis presenting with congestive cardiac failure in adulthood. Shackman (1963) described massive hepatic haemangiomas in a 35 year old adult, but this patient presented with ascites and cachexia and was initially thought to have a hepatoma. Her lesions are congenital, but they did not produce symptoms until the age of 33 years and also only during the third trimester of pregnancy. The cardiac failure is obviously due to high cardiac output failure resulting from the massive hepatic arteriovenous shunting and was aggravated by the increased blood volume due to pregnancy. The presence of cardiomegaly, pulmonary plethora, systemic desaturation support this cause of heart failure.

She has improved with supportive medical therapy and it is unlikely that steroids, radiation therapy or surgery would be of any added value. The large collateral blood supply from the splenic and intercostal arteries may indicate that hepatic artery ligation will not help to reduce hepatic arterial blood flow.

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