Cutaneous and hepatic haemangiomata

Haemangiomata of the skin in infancy are usually benign lesions which regress spontaneously. Hepatic haemangiomata or haemangioendotheliomata occasionally accompany multiple cutaneous naevi. The hepatic lesions may form arteriovenous fistulae, which when numerous give rise to high output congestive cardiac failure because of the considerable shunting involved. We describe the clinical course of multinodular hepatic haemangiomatosis associated with multiple cutaneous haemangiomata in a young infant, and the methods of treatment.

Case report

The patient was a 3-month-old girl, the second child of healthy, unrelated Greek Cypriot parents. Their first child had died in infancy, severely retarded after profound shock at birth caused by fetal haemorrhage from ruptured vasa praevia. This second baby was referred to the paediatric clinic by her practitioner because of parental concern over multiple skin naevi, some of which had been apparent shortly after birth. Her progress otherwise had been considered normal. She was a lively, well looking, if slightly pale infant whose weight was on the 50th centile (birthweight 9th centile). She had a left convergent strabismus. A total of 9 small cavernous haemangiomata, varying from 1 to 5 mm in diameter, were scattered over the scalp, trunk, and lower limbs. Further examination showed a grade II pansystolic murmur to the left of the sternum, and an easily palpable liver 2.5 cm below the costal margin. A very tentative clinical diagnosis of haemangiomatosis of the liver in association with multiple cutaneous haemangiomata was made at this stage, but as the child seemed asymptomatic, her progress was followed carefully as an outpatient.

Over the next 4 weeks, however, the liver enlarged further to 5.0 cm, and the spleen became palpable 2.0 cm below the costal margin; there was a hyperdynamic cardiac impulse, the cardiac murmur increased in intensity, and she developed tachycardia with a gallop rhythm. The jugular venous pressure, however, was not raised; she had no detectable oedema, no dyspnoea, and there were no adventitial sounds in the lungs. She was admitted for investigation and treatment when just over 4 months old.

Investigations. Haemoglobin was 93 g/dl, serum iron 42 μmol/l (235 μg/100 ml), and platelet count normal. Electrocardiogram showed biventricular hypertrophy; chest x-ray confirmed considerable cardiomegaly and suggested pulmonary plethora. Cardiac catheterization and cineangiography (Dr. K. Hallidie-Smith) did not show any abnormality of the heart, major vessels, or venous pulmonary return; there was no intracardiac shunt, but the cardiac output was high (8.4 l/min per m²). Coeliac arteriogram (K.H.S.) showed a grossly abnormal vascular pattern in the liver, with dilatation of the intrahepatic branches of the hepatic artery, very large hepatic veins, and a very fast circulation time through the liver, appearances compatible with extensive arteriovenous communications within that organ. Thus a diagnosis of haemangiomatosis of the liver appeared to be substantiated.

Treatment and progress. The infant was digitalized but despite this her condition became less satisfactory. There was further enlargement of the liver (6.0 cm) and a hepatic bruit was heard for the first time. Weight gain, the rate of which had fallen steadily since she was first seen, stopped. At the age of 6 months a 4-week course of prednisolone (10 mg twice daily initially) was given, and at the end of it there had been a marked reduction in liver size (to 1.5 cm), the spleen was only just palpable, the hepatic bruit was no longer audible, and she gained weight (see Fig.). She was then maintained on digoxin alone, but the cardiac murmur and cardiomegaly persisted, and gradually during the next 8 weeks the liver enlarged to its former size, the spleen became easily palpable, and she failed to thrive satisfactorily, weight having fallen to the 3rd centile. A second and similar course of prednisolone was started at age 9 months, with an almost identical result; and once again after it had stopped there was a gradual increase in liver size, though never to its previous maximum. Digoxin was stopped at the age of 11 months, and just before she was a year the murmur lessened in intensity, a gradual reduction in liver size started and she began to grow at an increased rate. By the age of 18 months the murmur had disappeared, the liver edge was just palpable in the costal angle, the spleen could not be felt, the cutaneous haemangiomata had regressed almost entirely, electrocardiogram was normal, and chest x-ray showed a striking decrease in heart size. Her weight had increased to between the 25th and 50th centiles, and her developmental progress seemed normal.

Discussion

Multinodular haemangiomatosis of the liver is a rare but important entity in infancy causing high output cardiac failure secondary to shunting through arteriovenous fistulae. It is accompanied by striking hepatomegaly, and often though not invariably by
multiple cutaneous haemangiomata. The combination of this marked hepatomegaly, cardiomegaly, and cardiac murmur with an absence at least initially of raised jugular venous pressure, dyspnoea, and adventitial sounds in the lungs differentiates from cardiac failure due to ventricular septal defect with which it is most likely to be confused. The condition does not invariably present in this way, for Sarde-
mann and Tystrup (1974) described 2 infants in whom an obstructive jaundice persisted for several weeks after birth in association with multiple cutaneous and placental haemangiomata, but apparently without cardiomegaly. The skin haemangio-
mata regressed from the relatively early age of 5 weeks, as did the jaundice, and the authors supposed that haemangiomata had also been present in the liver, obstructing the main bile ducts. The prognosis is clearly not always as gloomy as the early reports suggest. Kundstader (1933) reviewed 15 fatal cases, and McLean et al. (1972) reported 3 of their own cases and reviewed 28 recorded in the inter-
vening period, the mortality in these 31 infants being 71 per cent. Braun et al. (1975) however discussed 7 cases, 6 of whom had survived over a follow-up period of 2 to 11 years.

Several approaches to treatment have been report-
ed. When the haemangiomata was confined to one area of the liver, resection was undertaken (Leonidas et al., 1973); where the lesion has been more diffuse, others have ligated the hepatic artery in an attempt to reduce shunting (deLormier et al., 1967). These surgical measures however are not without risk (see review by Touloukian, 1970). The place of radiotherapy was discussed by McLean et al., (1972), who concluded that results at present did not support its use. Steroids are thought to have caused striking regression in some extensive cuta-
neous haemangiomata (Fost and Esterly, 1968), though the exact mode of action is uncertain, and it was inevitable that they should be used to control the diffuse liver lesions and so reduce shunting (Touloukian, 1970). In our case regression of liver size during and for a short period after treatment with prednisolone was striking, but the improvement was short lived and it cannot be claimed that this therapy radically altered the natural history of cavernous haemangiomata to regress (Lister, 1938). We would nevertheless agree with Braun et al. (1975) that steroid therapy in combination with digitalis is the initial treatment of choice, and may control symptoms in the seriously ill infant until such time as spontaneous regression of the lesions occurs.

Summary

A 3-month-old infant presented with multiple cuta-
neous haemangiomata, and was found to have a cardiac murmur and hepatomegaly. The latter increased strikingly over the next 3 months, and was accompanied by cardiomegaly. Other usual signs of congestive cardiac failure were not present, however, and an arteriogram showed the presence of a grossly abnormal vascular pattern in the liver, confirming the clinical suspicions of hepatic haemangiomatosis. Two short courses of steroid therapy resulted in a marked but temporary decrease in liver size. After the age of one year, however, there was gradual regression of all abnormal clinical signs without further therapy.

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References


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Powder aspiration in children

Report of two cases

Acute aspiration of zinc or talc powder is a very dangerous condition in childhood, and several fatalities have been reported. Recently we saw 2 cases. The first showed the classical course with delayed respiratory distress, and extreme measures were needed to save the baby. Using an aggressive therapeutic approach we were able to avoid acute sequelae in the second patient. We report our experiences and give clinical recommendations.

Case reports

Case 1. A 7½-month-old girl was admitted to hospital 6 hours after aspiration of Fissan Baby powder (containing talc, zinc oxide, and other substances).

The child was found choking, her face covered with powder. The mother cleaned the upper airways, and normal breathing returned. About 2½ hours after the accident she developed steadily increasing respiratory distress and was admitted to hospital.

Physical examination (6 hours after the accident) showed a well-nourished and well-developed baby in severe respiratory distress (pulse 164/min, respiration 64/min, temperature 38°C). The upper airways were clear. Expiration was prolonged with scarce fine rhonchi over both lungs which were hyper-resonant to percussion. The liver could be felt 2 cm below the costal margin.

Laboratory results showed a capillary PCO₂ of 42 mmHg and a leucocytosis of 15 400/mm³ (15.4 × 10⁹/l) with a strong shift to the left. The first x-ray film of the chest showed fine and diffuse alveolar infiltrates on both sides, partially confluent, a barely visible interlobar fissure on the right, and a small pleural effusion on the left (Fig. 1).

Treatment was begun with intravenous infusions, ampicillin, cloxacillin, and prednisone. During the next 3 hours respiratory distress increased, the PCO₂ rising to 58 mmHg requiring nasal intubation and controlled ventilation under neuromuscular blockade with pancuronium (Loosco Baby Respirator, IPNV with maximal inspiratory pressure). Repeated vigorous bronchial washing with normal saline still showed, even after many hours, viscous whitish material. Besides the antibiotics and prednisone, aminophylline, orciprenaline, and potassium iodide were administered. However, it was impossible to ventilate the child sufficiently because of

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Fig. 1 Case 1. Chest x-ray 7 hours after powder aspiration. Note the alveolar infiltrates in both lungs with partial confluence, and the pleural effusion on the left.