Generalised lymphangiomatosis with chylothorax

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SUMMARY A 9 month old boy presented with acute respiratory distress and was found to have a left pleural effusion. The chylous nature of the effusion, multiple bony lytic lesions, and splenic cysts lead to the diagnosis of congenital lymphangiomatosis with chylothorax. Surgical intervention including pleurectomy was required after unsuccessful conservative management.

Chylothorax is rare in children. More than half of the cases occur as a complication of cardiothoracic surgery. It has been observed as a benign self limiting phenomenon in neonates. Chest trauma and thoracic malignancy account for most of the remaining cases. A case of chylothorax with lytic bony lesions due to lymphangiomatosis is described.

Case report

A 9 month old white boy presented with a three day history of tachypnoea. He was one of dizygotic twins born by caesarean section at 38 weeks' gestation weighing 3000 g. He was breast fed initially and was on solids and artificial milk at presentation. Family history was non-contributory.

On examination, although tachypnoeic at 72 breaths/minute with subcostal recession and nasal flaring, he was playful. He was pink in air and not feverish. A carinatum deformity was observed. Examination suggested a large left pleural effusion with contralateral mediastinal shift. Small bluish macules were observed on the back.

Chest radiography (fig 1) confirmed a large left sided effusion and revealed lytic lesions in the head of the left humerus and anterior end of several ribs. Skeletal survey showed lytic lesions in the left femur (fig 2). Sonography confirmed the pleural effusion and showed multiple cystic lesions in a slightly enlarged spleen. A computed tomogram of the chest did not add further information and a bone scan was normal. The liver was normal and there were no other features of abdominal malignancy or lymphadenopathy.

Initial blood count and concentrations of serum urea and electrolytes were normal and blood and urine cultures sterile. Total serum protein concentration was 46 g/l with serum albumin of 26 g/l. Iliac crest marrow aspiration and bone trephine were normal. Thoracocentesis relieved the respiratory distress and was followed by dramatic resolution of clinical signs. It yielded 330 ml of turbid pink fluid which was subsequently identified as chyle (protein 45 g/l, cholesterol 2-6 mmol/l, and triglyceride 8-8 mol/l). Microscopy showed white cells in abundance, 98% of which were mature lymphocytes. Culture was sterile.

Rapid reaccumulation of the effusion necessitated continuous pleural drainage. A low fat medium chain triglyceride diet was commenced. Surgical evaluation was felt necessary due to a continuous loss of 300 ml of chyle daily requiring multiple plasma and albumin transfusions. A loss of one kilogram in weight, hyponatraemia, hypoalbuminaemia, and lymphopaenia had supervened in the two weeks since presentation. At thoracotomy a diffuse welling of chyle was observed from the anterior and superior mediastinum as well as from both the thoracic duct and an enlarged accessory duct. Both ducts were ligated and pleurectomy performed.

On histological examination a specimen of the pleural tissue and from a bone biopsy showed

Fig 1 Chest radiograph of patient.
Discussion

Berberich et al described a 3 year old child with bilateral chyloous pleural effusions, multiple bone lytic lesions, and pathological fracture of the tibia. They reviewed five previously reported cases with congenital lymphangiomatosis of bone with chylothorax. Since then three other cases have been reported. The age at presentation ranged between 9 months and 17 years. The most common presenting symptom was respiratory distress. Four of the nine cases presented with bilateral pleural effusions. Four patients developed right sided chylothorax after initial presentation with a left effusion, and in one patient the chylothorax was confined to the left pleural cavity. In all cases bony lytic lesions were an incidental finding. All but one of these cases occurred in boys. Two patients developed pathological fractures, and one had associated chylopericardium. In no previous report was splenic involvement diagnosed ante-mortem, although most of the cases in the literature date from the presonographic era. In the current case, however, it was the detection of multiple 'splenic cysts' and the presence of bony lytic lesions that led to a radiological diagnosis of lymphangiomatosis.

Management of these patients remains speculative but it is notable that both cases managed conservatively died whereas only one of the seven managed surgically succumbed. It has been concluded recently that conservative management in the form of chest drainage and low fat diet is successful in most cases of childhood chylothorax. Most of the cases reviewed, however, followed cardiothoracic surgery. Conservative management usually involves a low fat intake supplement with medium chain triglycerides, which are absorbed directly into the portal venous system without forming chylomicrons. In one report the chylothorax resolved after intrapleural instillation of 50% dextrose. In another case reviewed the chylothorax was apparently due to an intrathoracic collection of lymph from a rib lesion and resolved after resection of the rib.

This review of cases of chylothorax with lymphangiomatosis lends support to a recent suggestion that when there is evidence of an underlying lymphatic disorder, this is an indication for earlier rather than later surgical intervention. Surgery usually involves ligation of thoracic duct and accessory lymphatic channels, thoracotomy, and lung decortication. We acknowledge the help of Dr M Liberman and Dr R Dinwiddie with the management of this case and the preparation of this report.
Multiple nerve palsies in β thalassaemia major

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SUMMARY  A patient with β thalassaemia major is described who developed a lower motor neurone facial nerve palsy on the left side, together with a phrenic nerve palsy on the same side, during the course of the illness. This complication has not been reported before in haemoglobinopathies.

β thalassaemia major is the commonest haemoglobinopathy found in Sri Lanka. Neurological complications have been described in thalassaemia but these are mostly confined to spinal cord compression due to extramedullary haematoipoiesis and auditory nerve involvement.

Case report

An 11 year old Sinhalese girl, who developed pallor together with progressive abdominal distension during the latter half of infancy, was diagnosed as a case of β thalassaemia major after relevant haematological investigations. She was the youngest in a family of five children and the other siblings were in good health. There was no history of consanguninity and the father had died of ischaemic heart disease at 50 years of age. From late infancy onwards she received numerous blood transfusions at infrequent intervals due to poor compliance. The aim was to maintain the haemoglobin concentration above 80 g/l. Only a few doses of desferrioxamine were given to her because it was not readily available.

At the age of 7 years she developed a classical left lower motor neurone facial palsy that did not resolve with time. There was impairment of taste in the anterior part of tongue but no hyperacusis. There was no evidence of middle ear infection or mastoiditis. Audiometry was normal. She developed corneal scarring due to exposure keratitis and a tarsorrhaphy was performed to minimise further damage to the cornea. At 7½ years of age she was admitted with severe pallor and congestive cardiac failure. A chest radiograph showed an elevated left dome of the diaphragm. Fluoroscopic screening confirmed paralysis of the left dome. The rest of the neurological examination, including funduscopy, was normal. At 9 years of age, as her splenic size had increased to 15 cm below the left costal margin and the transfusion requirements had increased, a splenectomy was performed. Since then she has been on monthly prophylactic benzathine penicillin treatment.

At present both her height and weight are below the 3rd centiles, she is normotensive, and has the typical thalassaemic facies. Her liver span extends 13 cm below the right costal margin. The most recent investigations were as follows: haemoglobin concentration 85 g/l; white cell count 10-5x10^9/l with polymorphs 55%, lymphocytes 35%, monocytes 3%, eosinophils 5%, and basinphils 1%; reticulocyte count 0.4x10^12/l. The peripheral blood picture showed hypochromic and microcytic red cells, pronounced anisocytosis, normoblasts, hypersegmented neutrophils, and few macrocytes; platelet count 290x10^9/l; alkaline denaturation test positive; haemoglobin electrophoresis showed adult haemoglobin 15% and fetal haemoglobin 85%. Radiographs of the hand showed rectangular metacarpals with reticulation and a radiograph of the skull showed widening of diploe with ‘hair-on-end’ appearance. Radiographs and serial tomograms of the cervical and thoracic spine did not show any paravertebral soft tissue shadows. Consent for a myelogram was refused.
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Arch Dis Child 1989 64: 1058-1060
doi: 10.1136/adc.64.7.1058