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Primary Generalised Lymphatic Dysplasia

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The manifestations of primary lymphatic dysplasia in children occur as a consequence of congenital maldevelopment of the lymphatic systems. Often, only a single site is involved and the presentation is with lymphedema or chylous ascites or chylothorax. The rare combination of all 3 components in a single patient is termed as generalized primary lymphatic dysplasia and was first reported by McKendry *et al.*(1). Since then another eight authenticated case reports have appeared in the world literature(2,3). The rarity of the condition prompts us to report one such infant who, in addition to the other features, had a large chylopericardium.

Case Report

A 3-month-old boy, the first born in a non-consanguinous marriage, presented with gradually increasing abdominal distension since birth and fever for 2 days. Examination showed a febrile, anemic infant weighing 4 kg with tachypnea and bilateral pulmonary rales. In addition, he had non-pitting edema of the dorsum of the left hand and a soft, non-transilluminant swelling of the right side of tne neck. There was a tense ascites and bilateral reducible inguinal herniae (*Fig. 1*). Investigations revealed a hemoglobin of 6.2 g/dl and polymorphonuclear leucocytosis, with an absolute lymphocyte count of 1.7×10^9/L. *Staphylococcus aureus* was isolated from the blood culture. An *X*-ray of the chest showed normal cardiac size, bilateral bronchopneumonia and mild bilateral pleural effusions. The total serum protein was 5.2 g/dl with an albumin fraction of 2.0 g/dl. Thoracic and abdominal paracenteses drew acellular, sterile milky fluid. This fluid had a specific gravity of 1015, fluid:

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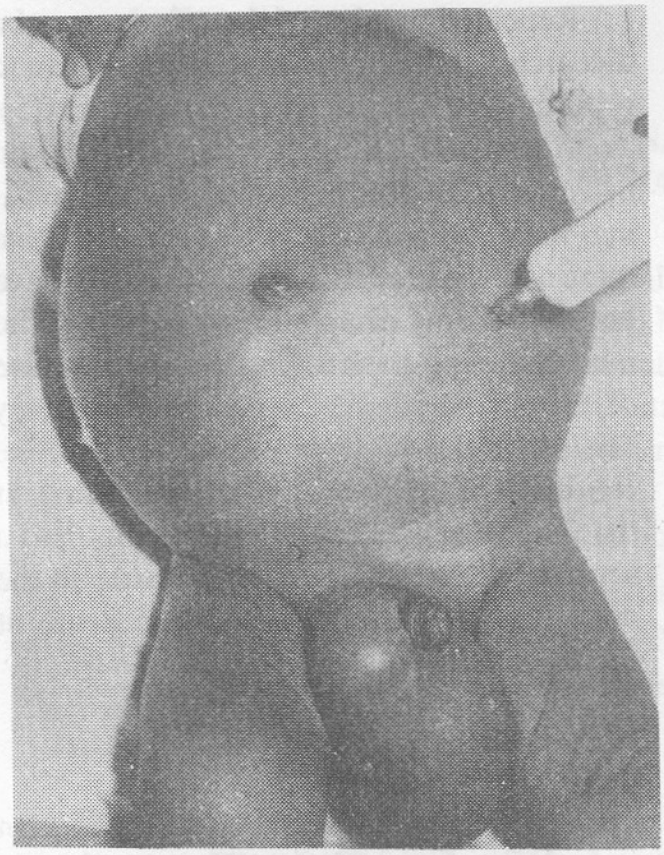

Fig. 1. Tense chylous ascites and bilateral inguinal herniae.

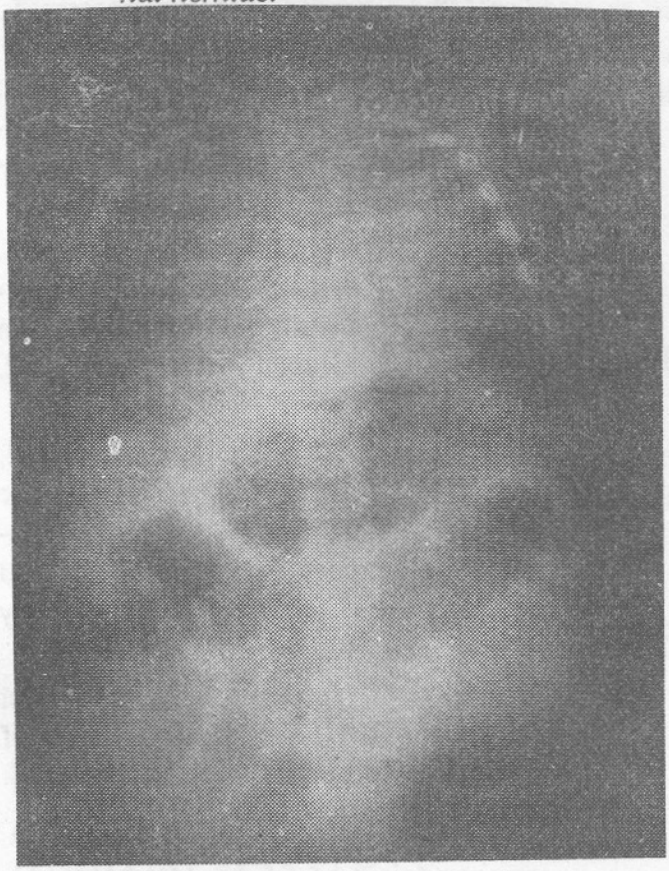

Fig. 2. X-ray chest during readmission showing a large mediastinal shadow.

serum ratios of protein, cholesterol and triglyceride were 0.92, 0.95 and 1.2, respectively. An ultrasound of the abdomen and thorax, an IVP and a barium meal study failed to identify any structural abnormality in the viscera.

Staphylococcal septicemias and bronchopneumonia responded well to antibiotics. A packed cell blood transfusion was given and therapeutic abdominal paracenteses were carried out thrice during the 3 week hospital stay. The parents were unable to afford total parenteral nutrition. Consequently, early feeding was instituted. The diet comprised of breast feeds and skimmed milk enriched with coconut oil and protein hydrolysates. Fat soluble vitamin were supplemented. There was symptomatic improvement. At the time of discharge, the child was afebrile and had minimal ascites.

The baby was readmitted 3 months later with fever and respiratory distress. Peripheral lymphedema was persisting, ascites had reaccumulated and there was a clinical evidence of bronchopneumonia. Chest *X*-ray showed a large mediastinal shadow (*Fig. 2*) and echocardiography confirmed the presence of a pericardial effusion. This was tapped twice, drawing 15 ml and 56 ml of milky, blood-tinged fluid. Subsequent attempts at tapping were unsuccessful. Radiological evidence of an abnormal mediastinal silhouette persisted. Blood and pericardial fluid were sterile on culture. Administration of broad-spectrum antibiotics resulted in cleaning of the pneumonic illness. However, about 3 weeks after hospitalization, there was sudden deterioration in the infant's condition and he died a few hours later. Permission for autopsy was not granted.

Discussion

The diagnosis of a generalized, primary lymphatic dysplasia in our case was based on the simultaneous occurrence of chylous ascites, lymphedema, bilateral chylothorax and chylopericardium. The disorder is extremely rare with only a handful of cases having been documented(2,3). The exact etiopathogenesis is not known. It is postulated to be due to either a lack of communicating channels between the smaller and larger lymphatic vessels(1) or to an obstruction and consequent leakage at the level of the cisterna chyli(2) or thoracic duct(4).

Only one of the 9 cases reported so far had a pericardial effusion(2). This was minimal and was demonstrated only at autopsy. Our case had a significant amount of pericardial effusion detected antemortem. A large mediastinal shadow persisted inspite of pericardiocentesis suggesting that there was also a mediastinal hygroma. This, however, could not be confirmed. In the case that has been already reported(2), the mediastinum contained chronically edematous tissue that leaked chyle when incised. Another unusual feature was the presence of bilateral inguinal herniae, a manifestation that has not been recorded in any of the reported cases. It probably resulted from increased intra-abdominal pressure caused by a tense chylous ascites.

Intestinal lymphangiectasia may be associated and may lead to a protein-losing enteropathy or a secondary immunodeficiency due to a loss of immunoglobulins and T-lymphocytes into the accumulated lymphatic fluid(2). The immune profile in our patient was not studied.

Repeated tapping of the effusions and a diet rich in medium chain triglycerides and protein form the mainstay of treatment(5). Total parenteral nutrition, surgical ligation of lymphatics, and peritoneovenous shunting have been tried where conservative treatment has failed(3,5). The results have been uniformly disappointing. Seven of the nine reported patients have died because of an overwhelming sepsis or respiratory failure caused by huge effusions(2,3). The condition of the two survivors was worse at the time of reporting(2). Our patient had a severe, generalized disease involving all the major serous cavities resulting in death at the age of 6 months.

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