Case Reports

Two children with chylothorax

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Introduction
Although chylothorax is the most common cause of pleural effusion in neonates, the incidence in the paediatric population is unknown[1,2]. We present 2 children who presented with chylothorax and responded to conservative management.

Case 1
A 2 month old male infant was admitted with cough for 4 days and respiratory distress for 1 day. The baby was afebrile with a respiratory rate (RR) of 84/min, a heart rate (HR) of 173/min and a saturation of 88% in room air. There was diminished air entry in the right side of the chest. Perinatal history was uneventful. There was no history of contact with tuberculosis. The baby did not have any dysmorphism. The chest x-ray (CXR) was suggestive of a right sided pleural effusion (Figure 1) which was confirmed by ultrasonography.

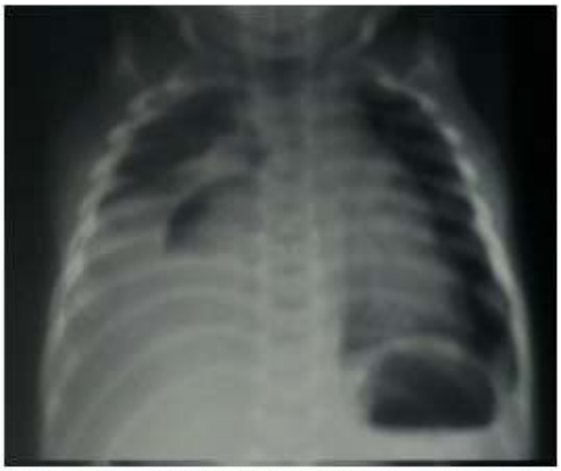

Figure 1: ***Right sided pleural effusion***

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An intercostal chest drain (ICD) was inserted and biochemical analysis confirmed the chylous nature of the fluid. Blood parameters are documented in Table 1. There was improvement in respiratory parameters over the next 48 hours. Baby was kept nil orally and started on parenteral nutrition (PN). It was planned to avoid breast milk and initiate feeds with medium chain triglycerides (MCT), after fluid output in ICD had decreased to less than 10ml/kg. Feeding was started after 5 days with MCT. However 4 days later he had tachypnoea, tachycardia and desaturations with CXR showing hydropneumothorax on the same side (Figure 2).

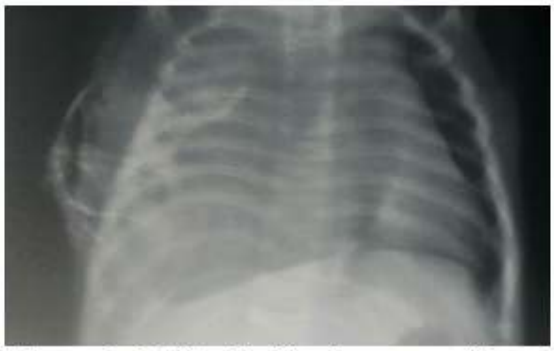

Figure 2: ***Right sided hydropneumothorax***

A repeat ICD insertion was done. There was significant clinical improvement thereafter. Feeds were re-introduced after 7 days and the chest drain was removed as there was no further accumulation of chyle. At the 6 months follow up, baby was growing well with a normal CXR at the last review.

Case 2
A 1 year and 10 month old girl, a known patient of critical pulmonary stenosis (PS) diagnosed at the age of 8 months, was admitted with breathing difficulty for 1 week. She was diagnosed to have tuberculosis on the basis of fever, pleural effusion and a Mantoux reading of 15mm, 4 months back. Initially she had been treated with multiple antibiotics with which the fever did not subside. However her fever subsided with improvement in serial CXRs after starting her on the 4 drug anti-tubercular therapy (ATT). On admission, she was afebrile with a RR of 74/min, HR of 112/min, a saturation 84% in room air and a mean blood pressure of 62mm of Hg. She had a loud systolic murmur over the left 2nd/3rd intercostal space. There was absent air entry on the left side of

chest. CXR was suggestive of pleural effusion (Figure 3).

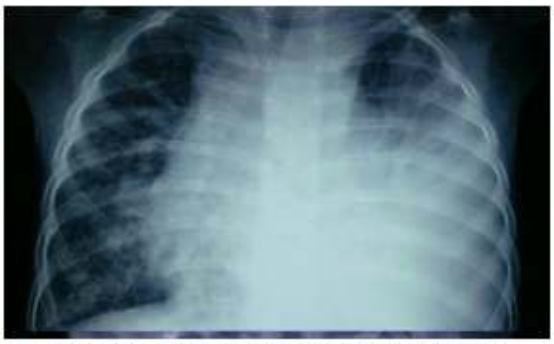

Figure 3: ***Left sided pleural effusion***

She had wide spaced eyes, short webbed neck, wide spaced nipples and her height was less than the 3rd centile for age and sex. She was presumed to be having Noonan syndrome on the basis of phenotypic features. She was on isoniazid and rifampicin for tuberculosis and propranolol and furosemide for her critical PS for which the compliance was poor. ICD insertion was done. Biochemical analysis was suggestive of chyle. She was kept nil orally. Blood parameters are documented in Table 1. There was clinical improvement over the next 72 hours following which she was started on a fat free diet. The ICD was removed after 7 days. ATT was continued for a total duration of 6 months. Genetic testing revealed mutation in PTPN11 which confirmed Noonan syndrome. She is now well in follow up and is due for cardiac surgery.

Table 1: ***Results of some investigations carried out in the two cases***

Investigations	Case 1	Case 2
Haemoglobin (g/dl)	10.7	8.1
Total leucocyte count (/cu mm)	12,500	7,800
Differential count (%)	N 72, L 24	N 56, L41
Erythrocyte sedimentation rate (mm)	09	21
Platelet count (/cu mm)	340,000	530,000
C-reactive protein (mg/L)	04	12
Alanine aminotransferase (U/L)	39	168
Mantoux test	Negative	Negative
Pleural fluid analysis		
Colour	Milky white	Milky white
Cell count	>15,000	>10,000
Differential count (%)	N 05, L 95	N 08, L 92
Glucose (mg/dl)	90.0	75.0
Protein (g/dl)	03.0	03.5
Albumin (g/dl)	02.0	02.8
Triglycerides (g/dl)	2760	2500
Lactate dehydrogenase (U/L)	300	410
Gene Xpert from pleural fluid	Negative	Negative
Gram stain, Ziehl-Neelsen stain and culture of pleural fluid	Negative	Negative

Both patients did not have any history of trauma, child abuse, vomiting or cough. CT thorax in both of them were suggestive of pleural effusion. There was no documentation of lymphadenopathy, vascular malformations or tumours.

Discussion

Management of chylothorax involves draining of the pleural fluid, diagnosis of aetiology, and prevention of malnutrition and immunodeficiency[1]. Surgical management should be attempted only if conservative management has been unsuccessful after 2-4 weeks. Initial management includes thoracocentesis for biochemical confirmation of chyle followed by drainage if needed by ICD. Ventilation with high positive end expiratory pressure may also help in sealing the thoracic duct. Medical management also involves using octreotide[3-7]. We inserted ICD in both our patients with chylothorax which helped in alleviating the respiratory symptoms. Conservative management has been shown to resolve more than 80% children with chylothorax[1,3,7].

In children, the common causes are congenital malformation of lymphatics, child abuse, trauma, post cardiac surgery or in association with syndromes (Down, Noonan and Turner). It can be associated with coughing, vomiting, subclavian vein catheterisation, tumours (neurogenic, lymphoma, teratoma) or infections (tuberculosis, histoplasmosis, sarcoidosis)[1,4,6,8]. The underlying aetiology should be evaluated with imaging studies (lymphography, lymphoscintigraphy, computed tomography, magnetic resonance lymphangiography, thoracoscopy). We could not ascertain the aetiology for our first case due to financial constraints but he is well in 6 months

follow up with no further recurrence of symptoms. The second child had Noonan syndrome with tuberculosis both of which are known to have an association with chylothorax.

Nutritional support with PN and enteral nutrition with fat free diet and MCT are recommended[1,2,9]. In resource limited settings where surgical facilities are not available, this can be an effective means of management. Patients are also prone to infections due to hypogammaglobulinaemia and lymphocyte depletion[10]. Both our children were kept nil orally till the drain output decreased to less than 10ml/kg which has been documented as a sign of good outcome in some studies[3]. PN was given during this period. Fat free diet with MCT was initiated thereafter. This reduces the intake of long chain fatty acids which are transported by chylomicrons and thereby decreases the leak through the lymphatic channels. Unfortunately, the first child had a hydropneumothorax possibly due to displacement of the ICD. MCT was continued for 3 months for case 1 and for 2 months for case 2 although there are no specific guidelines for the duration of continuation of MCT based diet. Conservative management with strict asepsis and proper nutritional support helped in treating both these patients in a set up where surgical facilities were not available.

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