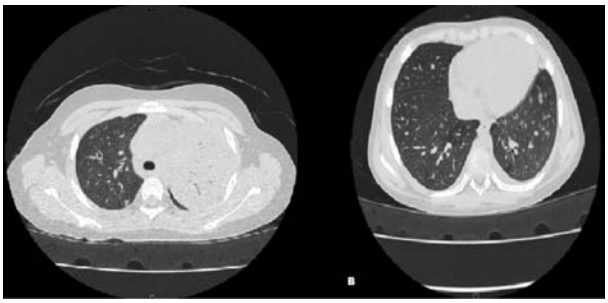

FIG. 2 *Image showing HRCT chest before (a) and one month after (b) induction chemotherapy.*

myeloid leukemia, but it may indicate leukemic transformation in myelodisplastic disorders, chronic myeloid leukemia, myelofibrosis, polycythemia vera or chronic eosinophilic leukemia [2,3]. Granulocytic sarcoma does not seem to have any prognostic significance in acute leukemia [4].

Granulocytic sarcoma in the lung is a rare entity [4]. In our case, the patient had an opacity visible in his chest *X*-ray and CT scan. Such focal masses during the course of acute myeloid leukemia may be an infection, hemorrhage or secondary neoplasms, apart from a granulocytic sarcoma [5]. It is known to be confused with opportunistic infections of the lung [6,7]. A diagnosis of granulocytic sarcoma is usually based on its appearance, location and a concurrent diagnosis of AML [8]. The tissue confirmation is done by morphology, and with stains like myeloperoxidase, periodic-acid schiff and neuron specific enolase [8]. The appearance of myeloblasts can range from well differentiated to poorly differentiated within a granulocytic sarcoma [9]. In our child, a possibility of fungal and bacterial infection was considered but BAL showed blast cells confirming the diagnosis of granulocytic sarcoma.

We conclude that a consolidation on chest radiograph in acute myeloid leukemia can be a granulocytic sarcoma of the lung; a bronchoalveolar lavage may be offered to confirm or refute this diagnosis.

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Skimmed Milk Preparation in Management of Congenital Chylothorax

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Background: Treatment for congenital chylothorax is based on adequate drainage of the pleural fluid and total parenteral nutrition followed by re-establishment of feeds using medium-chain-triglycerides based milk formulas which are expensive and not easily available. **Case characteristics**: Two newborns (one term and one preterm) with congenital chylothorax. **Intervention:** Skimmed milk preparation for enteral nutrition to provide high protein and low fat diet. **Outcome:** Successful resolution of chylothorax. **Message**: Skimmed milk preparation may be used for enteral nutrition of babies with congenital chylothorax where other feeding alternatives or commercial formulas are either not successful or are not available.

Keywords: *Chylothorax, Neonate, Octreotide, Skimmed milk preparation*

Chylothorax is a collection of lymphatic fluid within the pleural space, and is relatively uncommon [1]. Case fatality rate of congenital chylothorax, when complic2ated with hydrops, can reach upto 98% [2]. The diagnosis of chylothorax is made in the presence of white cell count of greater than 1,000 cell/mm^3 with predominance of mononuclear cells along with high triglyceride levels (≥1.2 mmol/L or 106.2 mg/dL) [3]. Medical management includes pleural drainage, cessation of oral feeds with total parenteral nutrition (TPN), diet modification with milk low in fat content, and octreotide. We report the successful use of skimmed milk preparation for enteral nutrition in congenital chylothorax.

CASE REPORT

Case 1: A female weighing 2030g was born at 33 weeks of gestation to a 29-year-old mother by elective caesarean section done in view of antenatally detected hydrops with increasing bilateral pleural effusion. There was history of two previous intrauterine deaths with fetal hydrops. This baby was depressed at birth with APGAR scores of 6 and 9 at 1 and 5 minutes, respectively. She had respiratory distress soon after birth, and was intubated and ventilated. *X*-ray showed bilateral pleural effusion; pleural tap done soon after birth drained 40 mL straw coloured fluid from right hemithorax, the analysis of which was suggestive of chylothorax (WBC count 4100/mm^3 with 98% lymphocytes, triglycerides 393 mg/dL, chylomicrons 400 mg/dL and protein 2.8 g/dL).

Chest tube was inserted on day 2 of life for progressively increasing fluid collection. She was started on gavage feeds with expressed breast milk (EBM) from day 3 of life, reaching 66 mL/kg/day by day 8 of life. In view of persistent pleural drainage, feeds were stopped and parenteral nutrition was started. She was also started on octreotide infusion starting at 1 µg/kg/hr from day 9 of life reaching 9 µg/kg/hr by day 14 of life. Trial of feeding with medium-chain triglyceride- based milk was unsuccessful. The dose of octreotide was kept almost constant *i.e.*, 9 µg/kg/hr until day 40 of life. Finally, skimmed milk preparation (prepared from skimmed milk powder (10g), table sugar (10g), coconut oil (2g) and water to make total volume 100 mL) was used for enteral nutrition in addition to intravenous octreotide from 32nd day of life. Pleural drainage started declining and finally ceased by 34th day of life. Baby was discharged on skimmed milk preparation along with fat soluble vitamin supplements on 42nd day of life. At 4 months, baby had reached a weight of 4000 g, and started on weaning foods. Her growth and development was appropriate for her corrected age on subsequent follow-up.

Case 2: A male baby weighing 3220 g was delivered at term to a 22-year-old mother by normal vaginal delivery. There was antenatal detection of a left sided pleural effusion. Baby was depressed at birth with APGAR scores of 6 and 9 at 1 and 5 minutes, respectively. Baby had mild respiratory distress soon after birth. Left sided chylothorax was diagnosed on biochemichal analysis (WBC 6200/mm^3, protein 3.4 g/dL, glucose 83 mg/dL, triglyceride 147 mg/dL, chylomicrons 102 mg/dL) of pleural fluid. He was started on gavage feeds using EBM soon after birth and graded up until 120 mL/kg/day by day 6 of life. Chest drain was inserted on day 6 of life in view of gradually increasing pleural fluid; feeds were stopped. He was started on TPN and kept nil-by-mouth from day 7 to day 16 of life. Skimmed milk preparation was started from day 16 of life. Feeds were graded up to 150 mL/kg/day by day 21 of life, and chest drain was removed on day 22 of life when there was no pleural drainage for preceding 5 days. He was discharged on skimmed milk preparation with no further re-accumulation of pleural fluid.

DISCUSSION

Congenital chylothorax may present with bilateral pleural effusions and respiratory insufûciency [2,4]. Hydrops may develop due to impaired venous return by cardiac and vena caval compression, and/or loss of protein into pleural space leading to generalized hypoproteinemia and edema [5]. Octreotide, a somatostatin analogue, has been used in various cases of congenital and postoperative chylothorax [5-7]. We unsuccessfully used octreotide infusion in the first baby. Special milk formulas [8] are not freely available in India in view of a stringent infant food act, and are far too expensive to use. Use of fat-free human milk was reported beneficial by Chan, *et al*. [9]. We, therefore, tried the formula prepared from skimmed milk powder which led to resolution of effusion in both babies. Adequate weight gain was documented during hospital stay, and after discharge, in both the children.

The use of pleural drain, TPN and octreotide is recommended in the initial management of congenital chylothorax. On resolution of the effusion, enteral feeds need to be started using low fat or fat-free milk formulas which are costly and are not freely available. Skimmed milk preparation with coconut oil (rich in medium-chain-triglyceride) could be a cheap and effective alternative to provide low fat and high protein calories in patients with congenital chylothorax where other feeding options have failed or commercial formulas are not accessible.

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Macrophage Activation Syndrome in Kawasaki Disease

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Background: Kawasaki disease is an acute febrile vasculitis of childhood. Macrophage activation syndrome is a rare life threatening complication. **Case characteristics:** 4-year-old boy with Kawasaki Disease treated with intravenous immunoglobulins. **Observation:** He developed encephalopathy, hepatosplenomegaly and pancytopenia. Blood investigations and bone marrow aspiration suggested macrophage activation syndrome. **Outcome:** Good response to pulse methylprednisolone (30 mg/kg/d) for 5 days. **Message:** Macrophage activation syndrome may complicate Kawasaki disease.

Keywords: *Lymphoproliferative disorders, Mucocutaneous lymph node syndrome.*

Macrophage activation syndrome (MAS) occurs secondary to many diseases, including infections, neoplasms, hematological conditions, and rheumatic disorders. It is characterized by persistent fever, pancytopenia, liver dysfunction, hepatosplenomegaly, hyperferritinemia, hypofibrinogenemia, elevated serum lactate dehydrogenase, and hypertriglyceridemia [1,2].

CASE REPORT

A 4-year-old boy was admitted with history of high fever for 14 days. He had a history of diffuse maculopapular truncal rash which started on day-4 of fever and persisted for 4 days. There was bilateral non-purulent conjunctivitis from the 3rd to 6th day of fever along with erythema of tongue and lips. Blood counts done elsewhere on day-12 were: hemoglobin 9.6 g/dL, total leukocyte count 18.8 × 10^9/L, platelet count 886 × 10^9/L, Erythrocyte sedimentation rate (ESR) 70 mm in 1st hour, and C-reactive-protein (CRP) 86 mg/L. Widal and Mantoux tests were negative. On examination, he was irritable, and had pedal edema, orange brown chromonychia, right cervical lymphadenopathy and hepatosplenomegaly. Investigations showed serum sodium of 130 mmol/L, Alanine aminotransferase (ALT) of 263 U/L, serum albumin of 2.8 g/dL, and a sterile blood culture. Urine microscopy revealed 10-12 pus cells/ high power field; culture was sterile. Echocardiography showed perivascular brightness with lack of tapering in left anterior descending artery and an aneurysm measuring 5 mm. Aneurysm (4.6 mm) was also present in left main coronary artery. A diagnosis of Kawasaki disease (KD) was made and intravenous immunoglobulins (IVIg) were administered at 2 g/kg over 24 hours.

After being afebrile for 48 hours, fever recurred on day-17. He became drowsy, developed gum bleeding and further increase in size of liver and spleen. Repeat blood counts showed hemoglobin 6.8 g/dL, total leukocyte count 4.6 × 10^9/L, platelet count 16 × 10^9/L, ESR 12 mm in 1st hour and CRP 256 mg/L. ALT increased to 468 U/L, International normalized ratio was 1.8 and activated partial thromboplastin time was 68 seconds. Persistent fever, encephalopathy, hepatosplenomegaly, deteriorating liver function and pancytopenia along with