A 4-year-old child with fever and persistent cough

Case report

The presenting patient was a 4-year-old female who was hospitalised with fever and cough, which had persisted for the previous 2 months. There had been no prior respiratory symptoms and the perinatal course had been uncomplicated. The patient was subsequently referred to the Dept of Pulmonary Medicine (Federal University of Rio de Janeiro, Rio de Janeiro, Brazil) after abnormal chest radiography (Figure 1). An echocardiogram demonstrated normal cardiac anatomy, although pericardial effusion was noted. On pericardicentesis, a turbid and hemorrhagic liquid was revealed, which had negative cultures for a number of common microorganisms. Pericardium biopsy proved inconclusive.

At rest, the patient’s respiratory rate was 20 breaths per minute, with an absence of dyspnoea or cyanosis. Bilateral basal rales were observed on auscultation. The systemic examination was otherwise normal.

Figure 1
Chest radiograph.

Task 1
Interpret the chest radiograph.

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CASE PRESENTATION

A 4-year-old child with fever and persistent cough

The patient was treated with antibiotics for common microorganisms, and diuretics. The pericardial effusion and cough improved, but the pulmonary infiltrates persisted.

The results of complementary examinations were as follows: negative reaction to tuberculin skin test; and sputum smear negative for acid-fast bacillus. The chest computed tomography (CT) scan that was taken at this time is shown in figure 2.

Answer 1
The chest radiograph revealed diffuse interstitial infiltrates, Kerley B lines and an enlarged cardiac shadow.

Due to the persistence of fever and pulmonary infiltrates, treatment for tuberculosis (TB), with rifampicin, isoniazid and pyrazinamide, was initiated empirically with fever improvement but no radiological improvement. After 5 months, chest radiography continued to show diffuse interstitial infiltrates, Kerley B lines and enlarged cardiac shadow. An echocardiogram demonstrated pericardial thickening and normal cardiac anatomy. Helical CT showed no vascular anomalies.

Answer 2
The CT scan showed parenchymal inhomogeneity with increased interstitial markings.

Task 2
Interpret the CT scan.

Task 3
At this point, what would you do?
Following the open-lung biopsy, histological examination showed extreme dilatation of subpleural lymph vessels, compatible with congenital pulmonary lymphangiectasis (CPL; figure 3).

Clinical course

One month later, left-sided pleural effusion was noted and the thoracentesis showed an opalescent liquid of pH 7.85, triglycerides 1,800 mg per dL, cholesterol 93%, and a cell count with a predominance of lymphocytes. Parenteral nutrition with lipids was initiated with a decrease in pleural effusion, but it subsequently re-accumulated and the patient underwent three more thoracenteses. Over the following 5 months, the patient was hospitalised on three occasions for respiratory distress due to recurrent chylothoraces. A thoracotomy revealed extreme dilatation of the lymphatic channels over the parietal pleura and a pleurodesis with 2% silver nitrate solution was performed.

The child was recommended a low-fat diet and remained asymptomatic for 3 years. After this uneventful period, the patient presented with body bruising. Her blood tests showed 109,000 platelets, activated partial thromboplastin time 1.37 (normal value <1.3), prothrombin time 1.27 (normal value <1.20), fibrinogen 103 (normal range 200–400), and bleeding time, α-dimers, factors assays (VIII, IX and XI) and von Willebrand studies were normal. After 3 months, the patient died aged 7 years due to massive haemoptysis in another hospital.

Discussion

Pulmonary lymphangiectasis is a rare condition characterised by dilatation of the pulmonary lymphatics [1]. It may be of two types: 1) CPL, probably by anomalous development of pulmonary lymphatics; and 2) acquired, secondary to obstruction of lymphatic drainage or to congenital cardiopathy with obstruction of venous return. The commonest form of CPL involves a premature onset, with respiratory discomfort in the neonatal period, which is generally fatal [2, 3].

NOONAN et al. [4] divided CPL into three groups as follows: 1) as part of generalised lymphangiectasis; 2) secondary to pulmonary venous obstruction; and 3) isolated. In the diffuse form of isolated CPL, the disease may involve all areas of the lungs, whereas in the localised form the lungs are only partially affected. The diffuse form is more frequent in newborns, with premature onset, and is often fatal. The patient who presented here matched the diagnosis of diffuse isolated CPL, since no other anomaly was presented and both lungs were affected in a diffuse manner. However, there was no premature onset and she had been asymptomatic until 4 years of age.

Three cases of late-onset diffuse isolated CPL that have been reported in the literature presented...
similar features, including coughing, diffuse reticulonodular infiltrate, pericardial effusion and chylothorax [5–7], although there was no evidence of cardiopathy or lymphatic obstruction. In the current case, the turbid and haemorrhagic pericardium effusion may be re-interpreted as chylum, with blood and apoptotic lymphocytes appearing as neutrophils.

Diagnosis is based upon lung biopsy and histological examinations. Lymphography is useful to exclude lymphatic obstruction, but, in the current authors’ hospital, this was difficult to perform. High-resolution CT (HRCT) is useful for defining parenchymal and interstitial disease and for carrying out a precise study of the airway. The typical findings of CPL on HRCT are thickening of the interstitium, with patchy ground-glass opacities. However, HRCT must be associated with clinical findings and cannot be used as the sole diagnostic mode because the image findings are unspecific.

Since the patient presented here with diffuse interstitial infiltrate, persistent fever and coughing, treatment for TB was initiated, similar to the case reported by Diógenes et al. [5], given the great prevalence of TB in Brazil.

Treatment of diffuse isolated CPL is palliative to prevent lymph discharge into serous cavities. Initially, the patient received medium-chain triglycerides and then parenteral nutrition with lipids, but chylothorax re-accumulated. Occurrence of refractory chylothorax is common in patients with congenital lymphatic malformations [8]. In this case, the chylothorax may have been precipitated by lung biopsy. The patient received chemical pleurodesis because conservative treatment failed and surgical intervention was required. The most common methods are pleurectomy and chemical pleurodesis with or without thoracic duct ligation [8].

In this case, the minor alterations in the blood examination did not justify the patient’s death from massive bleeding. She was being followed by a haematologist because it is likely that another disease was involved. However, she died during the investigation and no autopsy was performed. Due to the small number of reported cases, it would be premature to reach any conclusive prognosis. Of the four reported cases, including this one, three resulted in death, and the outcome after treatment was not reported in one. In contrast, Barker et al. [9] have suggested that the prognosis of CPL with post-neonatal onset is excellent. This divergence may show that the clinical behaviour and prognosis of CPL depend on the extent of pulmonary involvement of the lymphatic dilatations and/or to other still unknown factors.

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