

Exogenous lipid pneumonia in children: a disease to be reminded of

IZABEL ATHAYDE SALGADO¹, CHARLENE CAVALCANTE SANTOS², JOÃO VICTOR SALGADO³, PATRÍCIA CONSORTE FERRAZ⁴, DENISE MARIA HAIDAR⁵, HILDA ALMEIDA PEREIRA⁶

¹Specialist in Pediatrics; Pediatric Infectology Resident, Universidade Federal do Maranhão (UFMA), Parasitological and Infectious Diseases Service, Hospital Universitário Materno-infantil, UFMA, São Luís, MA, Brazil

²Specialist in Neonatology, UFMA, Neonatology Service, Hospital Universitário Materno-infantil, UFMA, São Luís, MA, Brazil

³MSc in Health Sciences, Universidade de Brasília (UnB); Clinical Analysis Service, Hospital Universitário, UFMA, São Luís, MA, Brazil

⁴Medicine Student, UFMA, Hospital Universitário Materno-infantil, São Luís, MA, Brazil

⁵MSc in Health Sciences, UFMA; Pediatric Pneumology Service, Hospital Universitário Materno-infantil, UFMA, São Luís, MA, Brazil

⁶Specialist in Neonatology, UFMA; General Pediatrics Service, Hospital Universitário Materno-infantil, UFMA, São Luís, MA, Brazil

Study conducted at Hospital Materno-infantil, Universidade Federal do Maranhão, São Luís, MA, Brazil

Correspondence to: Izabel Athayde Salgado – Rua das Limeiras, qd.D, casa 17, São Francisco, CEP: 65075-260, São Luís – MA, Brazil, Phone: +55 (98) 3235-2888 – Fax: +55 (98) 2109-8026 – izabel.athayde@yahoo.com.br

©2012 Elsevier Editora Ltda. All rights reserved.

BACKGROUND

Lipoid pneumonia (LP), also known as pulmonary steatosis or pulmonary lipoidosis, is an uncommon chronic inflammation of the lung parenchyma with interstitial involvement caused by the accumulation of oily material in the alveoli. Chronic constipation is a common symptom in pediatrics, and physicians often use mineral oil to treat intestinal constipation in children. The accidental aspiration of lipid formulations is the most frequent cause of exogenous lipid pneumonia in pediatrics¹.

The radiological feature of the lipoid pneumonia can mimic many other lung diseases, including infections and carcinoma. Diagnosis of the lipoid pneumonia is often delayed or missed due to the nonspecific clinical manifestation and radiological appearances¹. Given this scenario, a report on a patient with a history of chronic constipation and mineral oil ingestion is presented.

CASE REPORT

A four-month-old boy was admitted to Hospital Universitário Materno-infantil (Universidade Federal do Maranhão) presenting diarrhea (frequency of 10 evacuations a day with mucus, without blood), increasing of abdominal volume, fever, and vomiting for 10 days. The patient had a history of meconium ileus, evacuating for the first time within ten days. After the initial 10 days, the patient had persistent difficulty in evacuation. Due to these circumstances, his mother had been administering mineral oil.

In addition, he was presenting cough without secretion and tachypnea for two days. He was exclusively breastfed, and his scheduled vaccines were completed. At the physical exam he was slightly pale, with tachypnea (requiring 5 L/min of mask oxygen 60%), dehydration, cyanosis, and jaundice; although feverless. Pulmonary auscultation showed a rude vesicular breath sound with softer rhonchi. In the laboratory exams, leukocytosis (66.8% neutrophils, 26.0% lymphocytes), positive C-reactive protein, 8.85 g/dL of hemoglobin, and serum

protein of 4.1 g/dL (albumin: 2.9 g/dL and globulin: 1.2 g/dL) were found. Chest radiography showed an infiltrate in the apical regions in both lungs (Figure 1). The first diagnosis was community-acquired pneumonia. Thus, empiric antibiotic therapy was started with ampicillin gentamicin (5 mg/kg/day). Four days later, his intestinal presentation had improved, but he still presented crackles and rhonchi in the pulmonary auscultation. Thirteen days later, he started presenting febrile peaks and a new chest radiograph was requested, still showing opacities in the parenchymal of both lungs (Figure 1).

One week later, he was having daily febrile peaks despite the antibiotics. Requests were made for a chest computed tomography (CT); a gastric lavage for tuberculosis research (three samples), which came negative; and another hemogram and CRP, that were normal. CT showed extensive consolidations and geographic ground glass opacities in both lungs, without any hilar lymphadenopathy, volume loss, mass, or airway compression (Figure 2).

Bronchoscopy with bronchoalveolar lavage (BAL) was performed and 10 mL of an opalescent fluid were divided in three samples. The samples were centrifuged and then analyzed using hematoxylin and eosin staining.

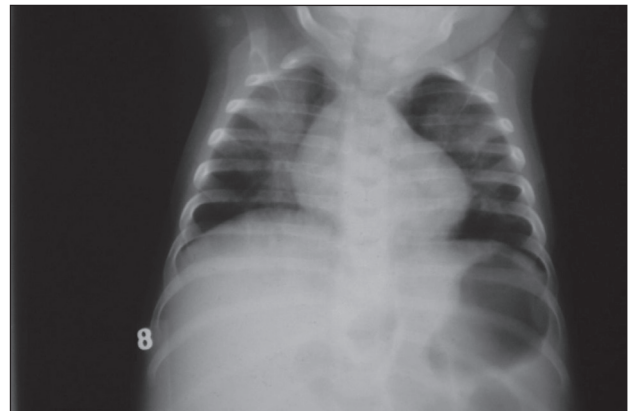


Figure 1 – Chest radiograph showing parenchymal opacities in apical regions both lungs.



Figure 2 – Computed tomography (CT) of the chest showing extensive consolidations and geographic ground glass opacities in both lungs.

The microscopic exam showed amorphous material, with inflammatory, red blood, cylindrical, and epithelial cells isolated and grouped. The exam also displayed frequent macrophages, some of them xanthomized and with lipid vacuoles (Figure 3). Thus, with the bronchoscopy, the diagnosis of LP was made. Our patient was in a stable condition and therefore discharged from hospital one month later, but with instructions to take prednisone 1 mg/kg/day for 30 days. Follow-up was conducted at the hospital pulmonology service. The follow-up chest radiographies showed a partial decrease of the parenchymal lesion, and the child was in good condition, without any clinical manifestations.

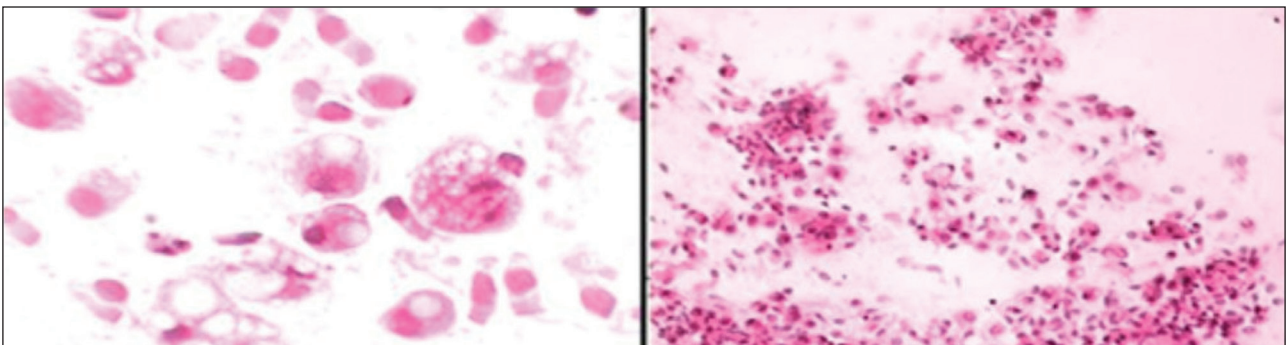


Figure 3 – Inflammatory reaction with lymphocytic infiltration collection of lipid-laden alveolar macrophages and free oil masses from the lung (hematoxylin and eosin staining/magnification, x 400).

DISCUSSION

Lipoid pneumonia can be classified as endogenous, idiopathic or exogenous. Endogenous LP, also called cholesterol pneumonitis, is less frequent and generally observed in patients with chronic bronchial obstruction of the airways by tumors, bronchiolitis obliterans, and lipid storage diseases such as Gaucher's disease and Niemann-Pick disease. Idiopathic LP is a rare disorder and has been associated with smoking in healthy individuals. Although exogenous LP is well documented in both pediatric and adult medical literature, it also occurs in the pediatric population as a rare condition caused by aspiration of mineral, vegetable, or animal oils. The use of mineral oil in the treatment of constipation or as an adjuvant treatment for partial bowel obstruction by *Ascaris lumbricoides* has been well accepted because of its efficacy and infrequent side effects¹⁻³.

In general, children present a widespread acute aspiration condition which progresses to respiratory failure and, sometimes, death. The presence of increasing dyspnea, cough, chest pain, fever, and vomit, together with alveolar infiltrates in the chest X-rays and the previous accidental ingestion of an oily substance, should be cause for suspicion of exogenous lipoid pneumonia. Moreover, the presence of oil in the lungs predisposes to recurrent infections, and in rare cases can be complicated by atypical mycobacterial and cryptococcal infection. These infections result in a bronchoalveolar macrophage response, followed by the release of inflammatory mediators, leading to fever, leukocytosis, neutrophilia, and elevated erythrocyte sedimentation rate. Due to its nonspecific clinical presentation and radiographic signs, exogenous LP can mimic other pulmonary diseases, such as bacterial pneumonia, tuberculosis, cystic fibrosis, bronchiectasis, and tumors^{2,4,5}.

Currently, analysis of the bronchoalveolar lavage (BAL) fluid is considered the diagnostic method of choice for suspected cases of LP, and only the cytochemical examination using staining can confirm the diagnosis⁶. Both

high resolution computed tomography (HRCT) and magnetic resonance imaging have showed suggestive signs of lipid infiltration of pulmonary parenchyma. However, HRCT is the best imaging modality for the diagnosis of exogenous LP^{5,7,8}.

In a recent review article, Marchiori et al.⁵ reported that there are still no studies in the literature that establish the best therapeutic option for LP. Nevertheless, the key procedure is interrupting the use of mineral oil, which leads to significant clinical improvement. Due to the fact that the depuration of the inhaled oil is a slow process, the best therapeutic strategy would be to withdraw the oil as soon as possible through bronchoscopy with multiple BALs⁶.

Gondouin et al.⁹, in a retrospective multicenter study, found that corticosteroid therapy has not been very effective for all cases of lipid pneumonia. The use of systemic corticosteroids in patients without clinical symptoms remains controversial. However, whole lung lavage, thoracoscopy with surgical debridement, and systemic corticosteroids have been used in patients with diffuse pulmonary damage. Corticosteroid therapy has also been suggested in cases in which functional and radiological alterations continue despite clinical improvement^{5,10}. Even though some retrospective case series demonstrated no benefit in the use of corticosteroids, the patient had a good response after this therapy, and remained asymptomatic at follow-up visits.

The present case emphasizes the need for increased awareness among caregivers and pediatricians about the potential hazards of mineral oil use for chronic constipation. Currently, despite no consensus on treatment, bronchoalveolar lavage seems to be an efficient therapeutic measure for the clearance of mineral oil from the lung parenchyma. This procedure may contribute to preventing fibrosis and reducing the morbidity of lipid pneumonia, which remains a rare diagnosis.

REFERENCES

1. Sias SM, Ferreira AS, Daltro PA, Caetano RL, Moreira JS, Quirico-Santos T. Evolution of exogenous lipid pneumonia in children: clinical aspects, radiological aspects and the role of bronchoalveolar lavage. *J Bras Pneumol*. 2009;35(9):839-45.
2. Khilnani GC, Hadda V. Lipoid pneumonia: an uncommon entity. *Indian J Med Sci*. 2009;63(10):474-80.
3. Hadda V, Khilnani GC. Lipoid pneumonia: an overview. *Expert Rev Respir Med*. 2010;4(6):799-807.
4. Schwaiblmair M, Berghaus T, Haeckel T, Wagner T, Scheidt W. Lipoid pneumonia - an underestimated syndrome. *Dtsch Med Wochenschr*. 2010;135(1-2):27-31.
5. Marchiori E, Zanetti G, Mano CM, Hochegger B. Exogenous lipid pneumonia. Clinical and radiological manifestations. *Respir Med*. 2011;105(5):659-66.
6. Sias SM, Daltro PA, Marchiori E, Ferreira AS, Caetano RL, Silva CS, et al. Clinic and radiological improvement of lipid pneumonia with multiple bronchoalveolar lavages. *Pediatr Pulmonol*. 2009;44(4):309-15.
7. Marchiori E, Zanetti G, Mano CM, Irion KL, Daltro PA, Hochegger B. Lipoid pneumonia in 53 patients after aspiration of mineral oil: comparison of high-resolution computed tomography findings in adults and children. *J Comput Assist Tomogr*. 2010;34(1):9-12.
8. Betancourt SL, Martinez-Jimenez S, Rossi SE, Truong MT, Carrillo J, Erasmus JJ. Lipoid pneumonia: spectrum of clinical and radiologic manifestations. *AJR Am J Roentgenol*. 2010;194(1):103-9.
9. Gondouin A, Manzoni P, Ranfaing E, Brun J, Cadranet J, Sadoun D, et al. Exogenous lipid pneumonia: a retrospective multicentre study of 44 cases in France. *Eur Respir J*. 1996;9(7):1463-9.
10. Russo R, Chiumello D, Cassani G, Maiocchi G, Gattinoni L. Case of exogenous lipid pneumonia: steroid therapy and lung lavage with an emulsifier. *Anesthesiology*. 2006;104(1):197-8.