Chylothorax in a Case of Accelerated Silicosis with Pulmonary Silicoproteinosis: A Unique Association

Archana Sasi, Animesh Ray, Ashu Seith Bhalla1, Sudheer Arava2, Shubham Agarwal, Ranveer Singh Jadon, Naval Kishore Vikram
Departments of Medicine, 1Radiodiagnosis and 2Pathology, All India Institute of Medical Sciences, New Delhi, India

Abstract

A 32-year-old gentleman, a worker in a cement-manufacturing facility with suspected silica-induced lung disease presented with acutely worsening Type 1 respiratory failure. With a negative work-up for infectious causes and no further revelations on bronchoalveolar lavage fluid or endobronchial biopsy, it was a transbronchial biopsy that ultimately led us to a diagnosis of silicoproteinosis with accelerated silicosis. Interestingly, the patient had a pleural effusion which on thoracentesis showed chylous fluid—the first reported case of chylothorax in association with silicosis.

Keywords: Accelerated silicosis, chylothorax, silicoproteinosis

INTRODUCTION

Silicosis is the most common occupational lung disease in the world. It is associated with a large number of vocations such as sand-blasting, cement-manufacturing and mining. Accelerated silicosis is a variant that develops within 5 to 10 years of exposure to silica dust. It is rare in comparison to chronic silicosis with single case reports or small case series reported in literature.1) Chylothorax is an unusual cause of pleural effusion that results from chyle leakage from the thoracic duct and contains a high triglyceride concentration. There is no previous report of chylothorax in association with silicosis.

CASE HISTORY

A 32-year-old male who had been working in a cement-manufacturing facility for 6 years (and had stopped a year ago due to illness) presented with complaints of dry cough for the past 1 year with progressively worsening shortness of breath over a period of 6 months. The shortness of breath was exertional and had progressed rapidly over the past 1 month. He also complained of low-grade fever every day for the past 1 month. There was no history of weight loss, loss of appetite, or of contact with tuberculosis.

On examination, he was found to be tachypneic with a respiratory rate of 30 breaths per minute and a pulse rate of 120 per minute with pulse oximetry revealing an oxygen saturation of 84%. His blood pressure was 112/76 mm Hg. Chest auscultation revealed fine bibasal crepitations. On presentation, he was anemic with hemoglobin of 11.9 mg/dL with normal leukocyte and platelet counts. His arterial blood gas report [Table 1] showed Type 1 respiratory failure. His chest X-ray was suggestive of bilateral diffuse infiltrates in the middle and lower zone. High-resolution computed tomography (HRCT) chest image revealed calcified mediastinal nodes with centrilobular nodules in upper lobes along with areas of interlobular septal thickening with fibrosis, a right-sided pleural effusion, and ground glass opacities seen in bilateral upper and lower lobes. [Figure 1a and 1b]

He was suspected to have a lower respiratory tract infection on a background of silica-induced lung disease. However, blood and sputum cultures were negative. He was empirically given multiple antibiotics along with noninvasive positive pressure ventilation.

On presentation, he was anemic with hemoglobin of 11.9 mg/dL with normal leukocyte and platelet counts. His arterial blood gas report [Table 1] showed Type 1 respiratory failure. His chest X-ray was suggestive of bilateral diffuse infiltrates in the middle and lower zone. High-resolution computed tomography (HRCT) chest image revealed calcified mediastinal nodes with centrilobular nodules in upper lobes along with areas of interlobular septal thickening with fibrosis, a right-sided pleural effusion, and ground glass opacities seen in bilateral upper and lower lobes. [Figure 1a and 1b]

He was suspected to have a lower respiratory tract infection on a background of silica-induced lung disease. However, blood and sputum cultures were negative. He was empirically given multiple antibiotics along with noninvasive positive pressure ventilation.

Address for correspondence: Dr. Animesh Ray,
Department of Medicine, All India Institute of Medical Sciences,
New Delhi - 110 029, India.
E-mail: doctoranimeshray@gmail.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Sasi A, Ray A, Bhalla AS, Arava S, Agarwal S, Jadon RS, et al. Chylothorax in a case of accelerated silicosis with pulmonary silicoproteinosis: A unique association. Indian J Occup Environ Med 2020;24:39-41.

Access this article online
pressure ventilation support. A fiberoptic bronchoscopy was done on high flow oxygen. The gross appearance of the bronchoalveolar lavage (BAL) fluid was normal and cultures sent for BAL fungal, bacterial, tubercular, and *Pneumocystis jirovecii* workup were negative. Endobronchial biopsy showed only exogenous pigment in the mucosal cells due to pneumoconiosis.

The patient then underwent a computed tomography (CT) guided-biopsy during which he developed worsening of shortness of breath, right-sided chest pain, and absent breath sounds on the same side. Chest X-ray showed a right-sided hydropneumothorax. A right-sided intercostal drain was inserted and this drained fluid which was opaque with a reddish hue [Figure 2]. Biochemical examination revealed an elevated triglyceride level of 898 mg/dL -suggestive of a chylothorax. A lymphangioscintigraphy was planned to identify the cause of the chylothorax but was deferred till stabilization of the patient’s condition. The patient’s condition initially improved after intercostal drainage with improvement in shortness of breath. However, he subsequently developed high-grade fever from the 5th day of CT-guided biopsy with worsening respiratory failure and new infiltrates on the chest X-ray. The patient also had new onset shock refractory to fluids and maximal inotropes. He was resuscitated as per the institutional protocol and antibiotics were added. Despite best efforts, the patient succumbed to septic shock due to the hospital-acquired pneumonia within 72 h. The transthoracic lung biopsy [Figure 3] showed revealed anthracofibrosis with alveolar proteinosis.

**DISCUSSION**

Silicosis is a lung disease that occurs due to inhalation and deposition of free crystalline silicon dioxide or silica in lung tissue. Accelerated silicosis generally demonstrates radiologic features similar to chronic silicosis with fibrotic changes on HRCT. However, features typical for silicoproteinosis may well be seen represented by bilateral air-space consolidation in the posterior dependent parts of the lung with calcified nodes. The case highlighted above appears to have developed accelerated silicosis with acute worsening due to development of silicoproteinosis.

BAL in silicoproteinosis typically yields a thick, milky effluent with periodic acid Schiff (PAS)-positive granular eosinophilic material and inflammatory cells similar to pulmonary alveolar proteinosis (PAP). For PAP, BAL is the principal diagnostic modality. However, a study by Briens *et al.* reported that 41% of patients did not have typical features on BAL. In a study by Inoue *et al.*, only 7.2% of patients required a transbronchial lung biopsy for a diagnosis of PAP, whereas combinations of HRCT picture, BAL, and transbronchial lung biopsy were adequate for the rest. A lung biopsy in silicoproteinosis shows intraalveolar accumulation of PAS-positive material.
Sasi, et al.: Chylothorax in a case of pulmonary silicoproteinosis

with peribronchial granulomatous inflammation. The lack of suggestive findings on BAL and transbronchial lung biopsy necessitated a CT-guided transthoracic lung biopsy.

We hypothesize that the cause of chylothorax in our case was lymphatic obstruction due to silicosis itself. Silica particles may disintegrate within macrophages in the lung and accumulate to cause a blockade of lymphatic outflow.[5] However, there have been no previous case reports describing the same. Chylous lymphatic leaks have been described as a complication of thoracic surgeries, primarily in operative procedures involving the esophagus, where major lymphatic ducts are damaged.[6] However, it has never been reported following transthoracic lung biopsy. Damage to the thoracic duct or associated structures seemed improbable as the site of biopsy was superficial. A lymphoscintigraphy might have revealed the site for leak or obstruction of lymphatic fluid; however, the general condition of our patient at the time did not allow for this investigation.

Treatment for silicoproteinosis is primarily supportive. Corticosteroids may provide short-term clinical improvement; however, they have not been shown to have any long-term benefit and increase the risk of infection. The role of whole lung lavage in its management is unclear. There have been reports of therapy success with this modality which is proven to be beneficial in pulmonary alveolar proteinosis.[7]

Our patient had pulmonary silicoproteinosis due to accelerated silicosis along with chylothorax likely due to extensive lymphatic obstruction due to silicosis. We believe that this is the first instance where a patient with pulmonary silicoproteinosis demonstrated opaque chylous fluid on thoracocentesis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Nakládalová M, Štepánek L, Kolek V, Žurková M, Tichý T. A case of accelerated silicosis. Occup Med Oxf Engl 2018;68:482-4.
2. Hutyrová B, Smolková P, Nakládalová M, Tichý T, Kolek V. Case of accelerated silicosis in a sandblaster. Ind Health 2015;53:178-83.
3. Briens E, Delaval P, Mairesse MP, Valeyre D, Wallaert B, Lazor R, et al. Pulmonary alveolar proteinosis. Rev Mal Respir 2002;166-82.
4. Inoue Y, Trapnell BC, Tazawa R, Arai T, Takada T, Hizawa N, et al. Characteristics of a large cohort of patients with autoimmune pulmonary alveolar proteinosis in Japan. Am J Respir Crit Care Med 2008;177:572-62.
5. Collis EL. Recent Views on Pneumoconioses. Proc R Soc Med 1931:531–42.
6. Hvass M, Fransen JL, Bruun JM. [Chylothorax]. Ugeskr Laeger 2017:179. pii: V05170429.
7. Stafford M, Cappa A, Weyant M, Lara A, Ellis J, Weitzel NS, et al. Treatment of acute silicoproteinosis by whole-lung lavage. Semin Cardiothorac Vasc Anesth 2013;17:152-9.

Figure 3: Photomicrograph of CT-guided lung biopsy shows: (a) Fibrosis with dense deposition of anthracotic pigment (anthracofibrosis). Fibrosis is highlighted by Massan trichrome stain (MT stain, blue color). (c and d) Special histochemical stain, periodic acid Schiff with diastase resistant (PAS D) reveals presence of granular proteinaceous material in the alveolar lumen suggesting alveolar proteinosis.