Pulmonary arterial dilation with normal pulmonary artery pressure in sarcoidosis

Anas Al-khateeb¹, William Meng², Muqueet Kadri¹, Sharath Bellary¹, Hari Sharma¹, Richard Miller¹

Departments of ¹Pulmonary and Critical Care and ²Internal Medicine, Saint Michael’s Medical Center, Newark, New Jersey, USA

Abstract

In this paper we present a clinical case that has improved on our knowledge and our curiosity about sarcoidosis. We report a case of a patient known to have pulmonary sarcoidosis, who presents with respiratory failure with severe hypercapnia. Following thorough investigations this patient was recognized to have three unique yet interrelated aspects of clinical manifestations. He was found to have severe bilateral diaphragmatic hypokinesis, dilated pulmonary vasculature with normal pulmonary pressure, and a state of high output right sided heart failure. We propose an explanation of such a presentation, while we attempted to discuss possible alternative mechanisms. In conclusion, we report this case as the first recognized case of sarcoidosis to be related to diffusely dilated pulmonary vasculature of normal vascular pressure.

Keywords: High-output heart failure, pulmonary arterial dilation, sarcoidosis

Introduction

Sarcoidosis has been associated with the development of pulmonary arterial dilation because of pulmonary hypertension. In this case, we report an unusual case of sarcoidosis with pulmonary arterial dilation without evidence of pulmonary hypertension.

Case Description

A 58-year-old African-American male, with past medical history of sarcoidosis, was presented for increased sleepiness and fatigue. Patient reported feeling shortness of breath for a few days, especially when lying down, but he mainly was brought to the hospital by his family because of excessive daytime sleepiness, which has been worsening gradually over the past few weeks. Reportedly, patient was diagnosed few years ago with sarcoidosis; diagnosis was made and biopsy was obtained via bronchoscopy. He reported a history of significant smoking about 25 packs/year and had been treated repeatedly as a case of exacerbation of obstructive lung disease. Patient quit smoking 2 years ago. On examination, vital signs were normal, except for respiratory rate of 24. He was lethargic but arousable to command. Breath sounds were decreased bilaterally, without wheezing, rhonchi, or rales. He had normal Jugular vein pulsation, normal heart sounds, without any audible murmur. He had +2 pitting edema on lower limb extremities. The rest of his examination was normal, including a BMI of 24.

Initial blood workup was significant only for microcytic anemia, low albumin, and metabolic alkalosis. Arterial blood gas confirmed our suspicion of respiratory acidosis and showed the following result: PH 7.28, PCO₂ 97, PO₂ 52, and SO₂ 90%. No A-a gradient was detected. Respiratory rate during this evaluation was 24. A chest X-ray done in the Emergency Department (ED) showed bilateral hilar masses with signs of basal atelectasis. Subsequently, we ordered fluoroscopic sniff test, which showed

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A chest CT scan was done to further evaluate the extent of his sarcoidosis and to rule out other illnesses [Figure 1]. It was significant for extensive generalized intrathoracic lymphadenopathy unchanged since 2 years. The Chest CT scan also showed moderate dependent atelectasis of both lung bases; and diffuse enlargement of the pulmonary vasculature. Measured diameter of main pulmonary artery, right main artery, left main artery, and ascending aorta were 5, 3.4, 2.8, and 4 cm, respectively. This was followed with transthoracic echo that showed normal ejection fraction of 55% with normal pulmonary artery pressure (PAP). Swan Ganz catheterization was performed and showed the following results: right atrial pressure 7 mmHg, right ventricular pressure 26/6 mmHg, PAP 28/12 mmHg, mean PAP 17 mmHg, and pulmonary capillary wedge pressure 5 mmHg. Cardiac output and index were 9 and 5.8, respectively. Oxygen saturation measured from right atrium, right ventricle, and pulmonary artery did not show any significant changes. A calibrated measured SVO2 was 88%. Further, blood workup was negative for collagen vascular disease, RPR, TSH, angiotensin converting enzyme, and schistosomiasis serology.

His clinical case was recognized as hypercapnic respiratory failure secondary to hypoventilation because of diaphragmatic muscle weakness as well as dilated pulmonary vasculature with normal PAP with signs of high-output right-sided heart failure. Patient responded well to noninvasive ventilation; he was discharged to subacute rehabilitation center to complete 4 weeks of physical therapy.

**Discussion**

This case has three interesting aspects of care, first of which was the most critical finding of severe respiratory hypercapnia. Although he was previously treated as a case of COPD, his A-a gradient was normal; this has led us to further investigate the etiology of his hypoventilation, which yielded a previously undiagnosed state of bilateral diaphragmatic weakness evidenced more with bilateral lower lobe atelectasis. Sarcoidosis has been reported as a cause of bilateral diaphragmatic weakness, either as sarcoid myopathy or as a result of neurosarcoidosis. Primary care physicians should be aware that home use of noninvasive ventilation after hospitalization with acute hypercapnic respiratory failure reduces mortality significantly.

Normal size of mean pulmonary artery is considered 29 mm in males and 27 mm in females, or depending on the pulmonary artery to ascending aorta ratio of less than 0.9. The most known association of dilated pulmonary artery is with pulmonary hypertension; however, it may be seen in a wide variety of other diseases that may cause dilated pulmonary artery without increasing the pulmonary arterial pressure. These have been categorized into increased blood flow (e.g., in cases of left-to-right shunting because of ASD), infectious, connective tissue disease, traumatic, and idiopathic cases. Rheumatologic and vasculitic diseases compromise the rest of the differential diagnosis, most typical of which is Behcet disease. The second interesting aspect in this case is that this is the first recognized case of sarcoidosis to be associated with pulmonary arterial dilation of normal pressure, after excluding other diseases known to cause pulmonary artery dilatation. The means by which we excluded other causes were extensively personal and family history, thorough physical examination, a panel of highly selected blood workup, and investigational studies of imaging and right heart cath. Although we cannot claim causality with this single report, future look out for this possible association is advised, especially in cases of sarcoidosis that are accompanied by hypercapnia. The main differential diagnosis in this case is what has been described as idiopathic pulmonary arterial dilatation, but it is less likely as the diagnostic criteria of which include the exclusion of chronic pulmonary diseases.

The third interesting aspect is his state of high-output right heart failure manifested with orthopnea and pedal edema, evidenced by high cardiac output and index, along with normal wedge pressure and normal lung parenchyma. Although the etiology of this is not entirely clear, we propose the theory that it is related to his severe hypercapnia. High-output heart failure has been reported as a possible cause of pulmonary arterial dilation because of pulmonary hypertension. Our case represents a different aspect of this spectrum, where it is possible the result of pulmonary arterial dilatation rather than a cause, given his normal pulmonary arterial pressure. We propose this theory after we have excluded other causes of high-output heart failure, such as sepsis, cirrhosis, hyperthyroidism, beriberi, and acquired Arteriovenous (AV) fistula. As for his anemia, his baseline hemoglobin was 9.4 g/dl and failed to show significant symptomatic improvement after the transfusion of one unit of RBC.

**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.

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Conflicts of interest
There are no conflicts of interest.

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