Case Report

Beyond Ortner's Syndrome – Unusual Pulmonary Complications of the Giant Left Atrium

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Abstract

Introduction: The giant left atrium (GLA) is a complication of severe mitral valve disease and causes morbidity by compressing adjacent intra-thoracic structures. Clinical Picture: We report 2 cases of unusual pulmonary complications of the GLA. Case 1 developed recurrent collapse of the left lung due to left main bronchus compression. Case 2 was diagnosed with right middle lobe compression and collapse. Treatment and Outcome: Case 1 was successfully treated by mitral valve replacement and left atrial reduction surgery. Case 2 was treated conservatively. Conclusion: Pulmonary atelectasis may occur in patients with GLA due to bronchopulmonary compression. Surgical management with valve replacement and atrial reduction may be necessary to relieve airway compression.

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Key words: Atelectasis, Cardiac surgical procedures, Heart atria, Mitral valve insufficiency

Introduction

The giant left atrium (GLA) is a complication of severe mitral regurgitation most commonly due to rheumatic heart disease. It causes morbidity by compressing adjacent intra-thoracic structures. Examples include Ortner's syndrome due to compression of the left recurrent laryngeal nerve, and dysphagia from oesophageal compression. We report 2 unusual cases of lung collapse due to bronchopulmonary compression by the GLA.

Case Report

Case 1

A 53-year-old man with severe mitral regurgitation from mitral valve prolapse presented to hospital in severe respiratory distress. He was intubated and transferred to the intensive care unit. The patient had several preceding hospital admissions for heart failure, and had become largely homebound due to worsening effort tolerance [New York Heart Association (NYHA) class IV]. He had refused cardiac surgery repeatedly, and had not been compliant to medical therapy. The baseline chest radiograph revealed cardiomegaly with left atrial enlargement (Fig. 1). Previous echocardiography showed a massively enlarged left atrium (13 x 12 cm), severe mitral and tricuspid regurgitation, pulmonary hypertension (pulmonary artery systolic pressure of 65 mm Hg), and a left ventricular ejection fraction (LVEF) of 50%.

The patient improved with diuretic therapy and was extubated 2 days later. However, he developed respiratory distress the following day and required re-intubation. Clinically, his trachea was deviated to the left, with dullness to percussion and diminished breath sounds on the same side. Chest radiographs showed left lung collapse that subsequently re-expanded with positive pressure ventilation (Fig. 2). He was weaned off the ventilator after another 3 days, but developed recurrent left lung collapse requiring intubation again. Bronchoscopy revealed severe extrinsic compression by the left atrium on the distal left main bronchus, resulting in near-total obstruction at the secondary carina (Fig. 3).

Following discussion with the family, the patient underwent mitral and tricuspid valve replacement surgery.

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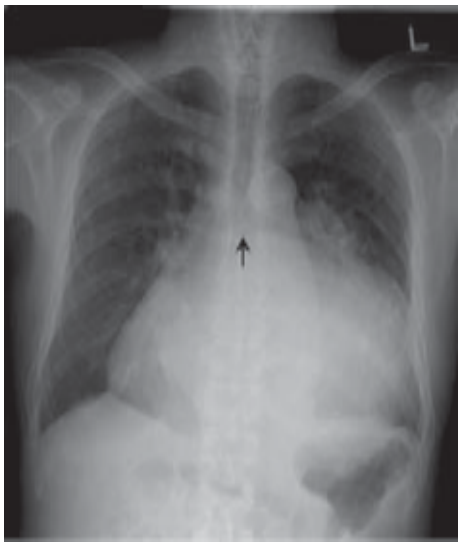

Fig. 1. Baseline chest radiograph of case 1 showing cardiomegaly (cardiothoracic ratio of 0.8), left atrial enlargement as evidenced by a prominent left atrial appendage, double right heart border, and splaying of the carina (arrow) with elevation of the left main bronchus.

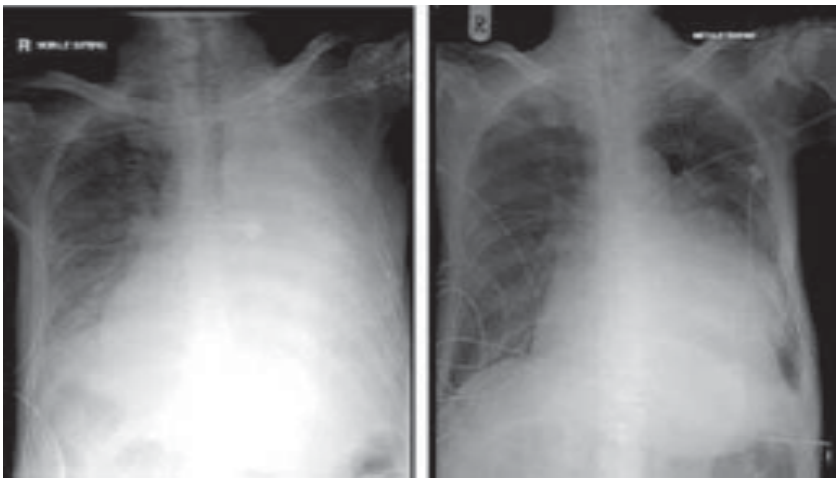

Fig. 2a. Mobile sitting.

Fig. 2b. Mobile supine.

Fig. 2. Chest radiograph showing left lung collapse that re-expanded with intubation and positive pressure ventilation.

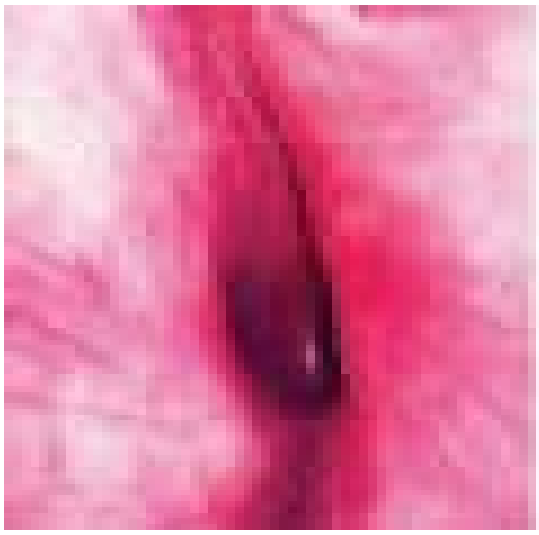

Fig. 3a. Preoperative bronchoscopy reveals extrinsic compression of the distal main bronchus and secondary carina.

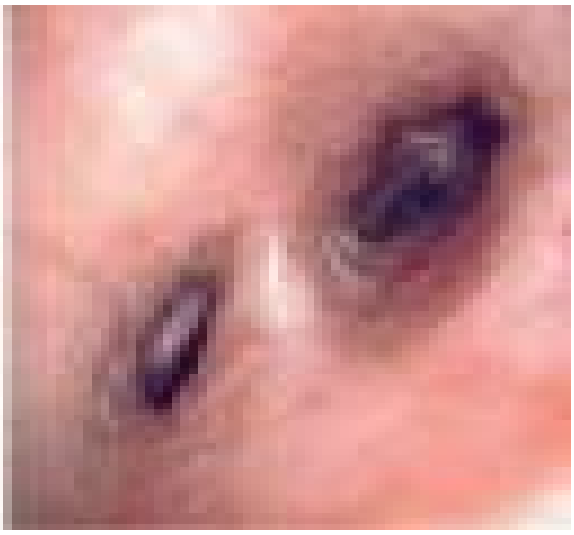

Fig. 3b. Postoperative bronchoscopy shows patent bronchi.

In addition, left atrial reduction surgery by plicating the posteroinferior wall was performed. There were no further episodes of atelectasis postoperatively. He was weaned off the ventilator and successfully rehabilitated. His functional status improved to NYHA class III. Repeat bronchoscopy 2 months post-surgery showed patent bronchi. Echocardiography showed reduction of the left atrial size from 13 x 12 cm to 7 x 5 cm.

Case 2

The second patient was a 67-year-old female who had undergone prosthetic mitral valve replacement 10 years ago for rheumatic heart disease. She had remained asymptomatic since the surgery, and her functional status was NYHA class II. Echocardiography showed an enlarged left atrium (9 x 7 cm), satisfactory prosthetic valve function, and a LVEF of 69%.

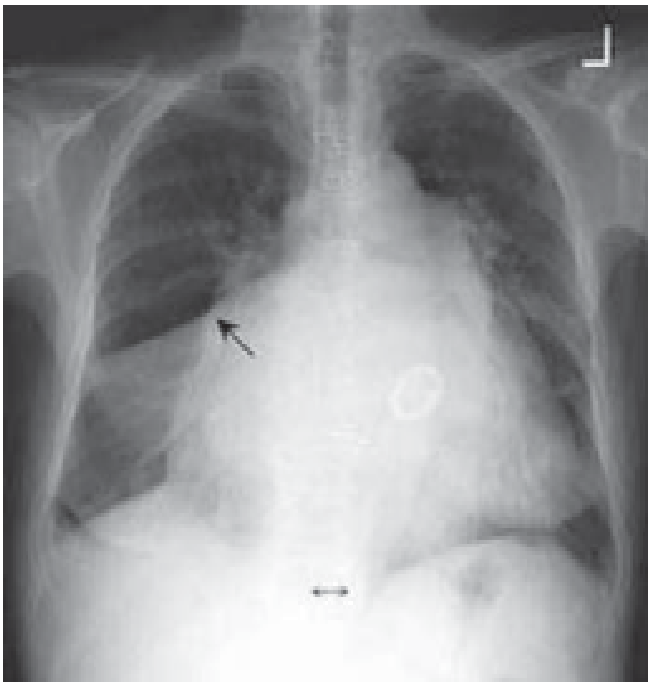

Fig. 4a. Chest radiograph of case 2 showing cardiomegaly (cardiothoracic ratio of 0.7), right middle lobe collapse as evidenced by depression of the horizontal fissure (arrow) and effacement of the right heart border. Left paravertebral tubular lucency due to laterally displaced oesophagus.

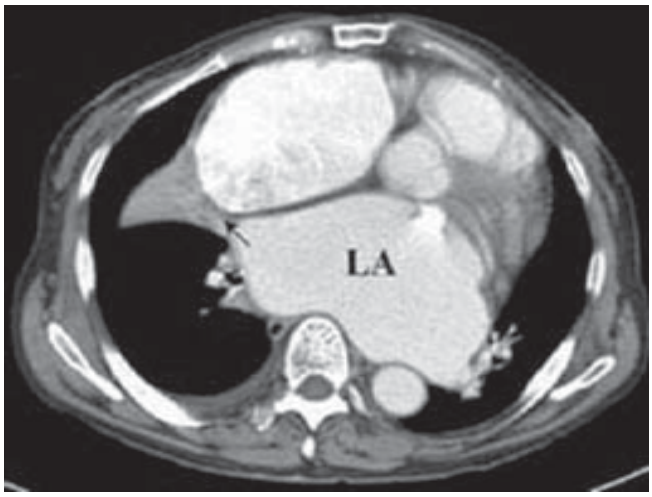

Fig. 4b. Computed tomographic scan showing left atrial enlargement (LA) and right middle lobe collapse (arrow).

She presented with right-sided pneumonia and was admitted for investigation. Chest radiograph showed cardiomegaly and collapse of the right middle lobe (Fig. 4a). Computed tomographic scan of the chest revealed severe enlargement of the left atrium (Fig. 4b). Bronchoscopy revealed extrinsic compression of the right middle lobe by the left atrium.

The patient was treated with a course of antibiotics and was discharged well from hospital. On subsequent follow-up, she remained asymptomatic and serial chest radiographs over 6 months showed no interval change of the right middle lobe collapse.

Discussion

The GLA was first reported by Owen and Fenton[1] in 1901 based on the post-mortem findings of a patient with rheumatic fever. Historically, the GLA was described as one where the left atrium touches the lateral chest wall on chest radiograph.[2] With the advent of echocardiography, it has been re-defined by several authors as a left atrium that measures more than 6 to 8 cm in diameter.[3,4] The GLA arises as a result of severe pressure and volume overload of the left atrium over a prolonged period of time. The underlying aetiology is usually rheumatic mitral regurgitation, and less commonly, mitral valve prolapse.[5]

In 1897, Ortner[6] described 2 patients with mitral stenosis and hoarseness of voice. This was then attributed to compression of the left recurrent laryngeal nerve between the enlarged left atrium and the arch of the aorta. In 1980, Morgan and Mourant[7] reported another complication of the enlarged left atrium, that of dysphagia due to compression of the oesophagus. To understand how these complications arise, one has to consider the anatomical position of the left atrium. Contrary to its name, the left atrium does not lie on the left side, but forms the most posterior chamber of the heart. It is closely related to the oesophagus, spine, left recurrent laryngeal nerve, pulmonary vessels, lung parenchyma and bronchi. When the left atrium enlarges, it causes complications by compressing these adjacent structures.

Pulmonary complications of the GLA are uncommon. Kawazoe et al[8] in 1983 described postoperative respiratory failure after mitral valve surgery in patients with GLA due to bronchopulmonary compression. However, to the best of our knowledge, de novo pulmonary atelectasis has not been reported previously. It is interesting that our 2 patients developed collapse of different lobes of the lung. We believe this is due to the enlargement of the atria in different directions. In the first case, leftward expansion of the left atrium resulted in left main bronchus compression. Conversely, compression of the right middle lobe was caused by rightward expansion of the left atrium in the second case.

The treatment of this condition consists of mitral valve replacement and left atrial reduction surgery to reduce atrial size and relieve compression on the bronchus.[8] We excluded airway stenting in case 1 as the compression

involved the bifurcation of the left main bronchus and was anatomically unsuitable for stenting. In addition, surgery had the advantage of correcting the underlying mitral valve disease and improving the cardiopulmonary status of the patient. In case 2, the patient was treated conservatively as the prosthetic valve function was satisfactory and she was otherwise asymptomatic.

In conclusion, pulmonary atelectasis may result from bronchopulmonary compression by the GLA. Therefore, in a patient with severe mitral valve disease and respiratory compromise, airway compression by the left atrium should be considered as a differential diagnosis. Surgical management with valve replacement and atrial reduction may be required to correct the underlying aetiology and relieve compression.

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