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ABSTRACT

Giant bullae are bullae that occupy at least 30 percent of a hemi thorax. This condition can rarely be idiopathic and not usually suspected in young patients with no risk factors. We describe a case of a giant solitary pulmonary bulla in a healthy young female with no known risk factors. 23-year-old female presented to the Emergency department with dyspnea and pleuritic right sided chest pain. She started experiencing these symptoms when she was on a 7-h flight and later experienced similar symptoms when she went scuba diving. Lung exam revealed decreased breath sounds on the right and she was saturating well on room air. Chest X-ray done showed a large bleb at the right lung apex. CT angiogram done was negative for pulmonary embolism, but confirmed a large bulla involving the right upper lobe. She had no history of lung diseases, marfanoid features, cigarette smoking, drug use or family history of similar condition. She underwent VAT assisted mini thoracotomy with resection of the right apical bulla and tube thoracostomy. Surgical pathology showed a pulmonary bleb with pleural fibrosis and prominent adhesions with parietal pleura and no evidence of malignancy. She was advised to avoid air travel and diving for 3 months and is doing well. Idiopathic giant pulmonary bullae have rarely been reported. It is a rare cause of dyspnea and chest pain in young adults. This may be suspected when patients develop symptoms with air travel and deep sea diving.

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1. Introduction

Giant bullae are bullae that occupy at least 30 percent of a hemi thorax. These usually develop because of cigarette smoking. Marijuana use, HIV infection, alpha 1 antitrypsin deficiency and intravenous drug use have also been described as possible causes [1,2]. Smaller bullae have been described in conditions like Marfan's syndrome, Ehlers-Danlos syndrome, Polyangiitis with granulomatosis and Sarcoidosis. This condition can rarely be idiopathic and not usually suspected in young patients with no risk factors as described above. We describe a case of a giant solitary pulmonary bulla in a healthy young female with no known risk factors.

2. Case description

23-year-old female who presented to the Emergency department with dyspnea and pleuritic right sided chest and upper back pain. She started experiencing these symptoms 8 days before presentation to the ER, when she was on a 7-h flight to Mexico. Symptoms subsided after she landed. She experienced similar symptoms when she went scuba diving there. Symptoms resolved when she surfaced, but she had a similar episode on the flight back from Mexico. The recurrent symptoms prompted her to come to the ER. She was saturating well on room air. Lung exam revealed decreased breath sounds on the right. Lab results for complete blood counts and basic metabolic profile were normal. EKG showed normal sinus rhythm and no ST or T wave changes. She had a chest X-ray done which showed a large bleb at the right lung apex, measuring approximately 14.5 \times 8.7 \times 12.2 cm. CT angiogram done was negative for pulmonary embolism, but confirmed a large bulla involving the right upper lobe and corresponding compression atelectasis of the right lung tissue without evidence of pneumothorax and normal left lung [Figs. 1 and 2]. Patient had no past medical history except for acute myelogenous leukemia at 3 years of age, for which she underwent chemotherapy without any radiation treatment. She had no history of lung diseases, marfanoid features, cigarette smoking, drug use or family history of similar condition. Alpha 1 antitrypsin levels were normal. Thoracic surgery was consulted. She initially underwent VAT assisted mini thoracotomy with resection of the right apical bulla and tube



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thoracostomy. Intraoperative bronchoscopy showed narrowing of right upper lobe apical segment with completely occluded airway segment which could not be traversed, secondary to extrinsic compression. As her lung did not completely re expand and showed a persistent right apical pneumothorax on the follow up chest Xrays, she underwent removal of old tube thoracostomy and a repeat chest tube insertion one week after the initial procedure. Surgical pathology showed a pulmonary bleb with pleural fibrosis and prominent adhesions with parietal pleura and no evidence of malignancy. Chest tube was eventually removed after 2 weeks as outpatient. She was advised to avoid air travel and diving for 3 months and is doing well.

3. Discussion

Idiopathic giant pulmonary bullae have rarely been reported. Patients may be asymptomatic, may experience dyspnea on exertion or rest, chest pain and occasionally present with hemoptysis, if acute hemorrhage into the bulla. These symptoms may be worsened with air travel and deep sea diving.

Boyle's law states that at a constant temperature, the volume of a gas varies inversely with the pressure to which it is subjected and Henry's law states that at a constant temperature, the amount of a gas that is dissolved in a liquid is directly proportional to the partial pressure of that gas. As a diver descends, the air in the lungs becomes compressed. Pulmonary edema and hemorrhage occur when lung volume decreases below residual volume. As a diver ascends and trans alveolar pressure exceeds 20–80 mmHg, overexpansion injury in the form of alveolar rupture can occur leading to pneumothorax and expansion of bulla [3]. Our patient experienced chest pain during the ascent of her dive.

As altitude increases, barometric pressure and atmospheric partial pressure of oxygen (PiO_2) both decrease. At 40,000 feet (12,192 m), the actual atmospheric pressure is 141 mmHg, whereas pressurization of the cabin on commercial airliners maintains cabin atmospheric pressures around 565 mmHg (at sea level, atmospheric pressure is 760 mmHg). Boyle's law states that the volume of a gas is inversely proportional to the pressure to which it is exposed. Thus, as barometric pressure falls in the aircraft cabin during the ascent, trapped air in any non-communicating body cavity (for example: non-communicating pneumothorax, lung bleb,



Fig. 1. Coronal section of CT thorax showing a large bulla involving the right upper lobe and corresponding compression atelectasis of the right lung tissue.



Fig. 2. Transverse section of CT thorax showing a large bulla involving the right upper lobe and corresponding compression atelectasis of the right lung tissue.

lung bulla, lung cyst, paranasal sinuses) will expand. Expansion of gas in the lungs is also increased by the high moisture content. Volume of air in a non-communicating body cavity can increase by approximately 38 percent upon ascent from sea level to the maximum "cabin altitude" of 8000 feet [4,5]. Our patient experienced chest pain during periods of ascent and turbulence during the flight.

The risk of a bulla or pneumothorax enlarging is increased if a patient flies less than 24 hours after diving, which was done by our patient. She may have had barotrauma during scuba diving and the flight less than a day later may have caused the bulla to increase in size based on the principles explained above.

Giant bullae are often detected when a chest X-ray is done to evaluate for dyspnea. CT thorax can confirm the diagnosis, examine the adjacent lung tissue and rule out a pneumothorax. Pulmonary function tests are done to assess airflow limitation. Case reports have described spontaneous regression of giant bullae [6]. Bullectomy is recommended for symptomatic patients [7].

Secondary pneumothorax is a common complication of giant bullae. Cases have also reported bronchogenic carcinoma arising within giant bullae [8]. Patients can also develop superimposed infection [9].

4. Conclusion

Idiopathic solitary pulmonary bulla is a rare cause of dyspnea and chest pain in young adults. This may be suspected when patients develop symptoms with air travel and deep sea diving.

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