Case Report

A Rare Case of Giant Pulmonary Bullae Compressing the Heart with a Brief Review

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Introduction: An air space of >1cm and air space occupying >1/3 of hemithorax are called as bullae and Giant Pulmonary Bullae (GPB), respectively. GPBs are rare and usually accompany smoking abuse-related diseases. The GPB may lead to hypoxia and or mass-like compression effect on neighboring healthy lung and mediastinum. The bullectomy is the gold standard but in surgically unfit cases one has to select result result-oriented, minimal-invasive procedure with considering risk and benefit.

Case report: A 27-year-old chest symptomatic female with idiopathic GPB occupying most of the right hemithorax with herniation to the contralateral side was detected on CT thorax with low cardiac ejection fraction on ECHO. A tremendously improved cardiac ejection fraction from 30 to 58% was achieved after a simple percutaneous intracavitary placement of a pigtail catheter with an underwater seal carried out with local anesthesia. Also, she could be weaned off from O_2 support.

Discussion: In a surgical unfit case of GPB the number of bullae, its size, site, and location (unilateral or bilateral) with evaluation of the underlying condition of lung and comorbidity is a prerequisite for intervention. An alternate minimally invasive procedure is to have opted to decompress the GPB. Spontaneous pneumothorax is the most common complication GPB, even without a percutaneous procedural approach, and can easily be managed with an underwater seal drainage tube.

Conclusion: The GPB may lead to chronic hypoxia and a mass-like effect, so to avoid life-threatening emergency situations decompression of GPB be considered followed by resection.

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Introduction

A bulla is defined as an air space in the lung parenchyma of more than one centimeter in diameter during full inspiratory state, while the term giant bulla is used when it occupies at least 30% of a hemithorax on chest X-ray. Idiopathic giant pulmonary bullae (GPB) is a rare condition with a predilection to male gender. Bilateral multiple bulla may accompany giant bulla as is commonly found with COPD or smoking abuse diseases. The other commonly used terminologies are vanishing lung/ MacLeod syndrome/ Bret syndrome/Swyer Swyer-James syndrome and bullous disease of the lung. The other etiologic factor includes cigarette and marijuana smoking, HIV infection, alpha-1 antitrypsin deficiency, and among intravascular drug users (methadone, methylphenidate, or talc-containing drugs etc) in Ehlers-Danlos type IV, polyangiitis with granulomatosis, Sjögren syndrome, and Sarcoidosis, but usually not a giant bullae.[1] The patient may be asymptomatic but most of them present with varying degrees of dyspnea along with exertion or at rest. Intra bullous infection with air-fluid level or hemorrhage may alter the clinical presentation but pneumothorax remains the most common complication. Radiologically at several occasions, it becomes difficult to distinguish GPB from Pneumothorax, because both have similar signs, symptoms, and radiological appearances so further imaging i.e. chest CT with contrast may be required for differential diagnosis.[2] However the double wall sign mentioned by Beatrice Araini et al is valuable to distinguish pneumothorax from adjacent giant bullae when both sides of the bulla wall is seen parallel to the chest wall.[3] A giant bulla is most likely to compress the surrounding healthy lung to cause hypoxia, chest pain or occasionally a mass-like effect on neighboring structures i.e. mediastinum, blood vessels or heart *(as also happened in our case).* These cases required further evaluation with computed contrast-enhanced

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CT thorax, ECG and 2 D ECHO, etc.[4] The mechanism of giant bulla formation is not very clear; it is usually the complication of emphysema with loss of elasticity in the wall of respiratory bronchioles or alveolar sac. The airway wall stretches and breaks down leaving behind a bigger air space with trapped air, which slowly over time increases in size, however sometimes the growth spurts suddenly and inflates to form a giant bulla. The check wall mechanism may play a significant role in these cases. In preexisting COPD patients at occasion with BiPAP and ventilator-associated management the bulla formation has also been reported due to positive airway pressure.[5] Morphologically the bulla remained separated from the lung parenchyma by a thin, fibrous, irregular membrane. The giant bulla are divided into four groups, which helps in management strategy for the selection of a procedure:[6]

Group I. Single giant bulla with underlying normal lung *(observed in our patient)*

Group II. Multiple giant bullae with underlying normal lung.

GroupIII. Multiple bullae with underlying lung broadly affected by emphysema.

Group IV. Multiple bullae with underlying lung affected by other diseases.

Limited interventional options are available for bullous lung disease. It depends on the number of bullae, its size, site, and location (unilateral or bilateral) with the underlying condition of lung or disease status. Bullectomy is preferred but for surgically unfit candidates, a minimally invasive endo bronchia valve (EBV) placement through a Bronchoscope or intra-cavitary drainage is the choice.[7] Endoscopic drainage of infected bulla is another option.[8] The percutaneous intra-cavity suction and drainage under local anesthesia was originally described by Monaldi (1938, 1947), for tuberculous cavities. John Alexander first time (in1946) applied Monaldi's procedure in bullous emphysema, followed by head and avery, 1949, since then it became a widely accepted technique. A historic aspect of other surgical procedures was mentioned by A. M. Macarthur et al that the use of plication (Benfield *et al*., 1966; Fitzgerald *et al*., 1974), local excision (Wesley *et al*., 1972), segmental resection (Head *et al*., 1960), lobectomy (Woo-Ming *et al*., 1963), and even lung transplantation (Veith *et al*., 1973), and in the recent past newer technique i.e., bulectomy, Vedio Assisted Thoracic Surgery (VATS), and Lung Volume Reduction Surgery (LVRS) etc become popular but each one had wide variable mortality.[9] Pneumothorax is the most common complication of the transcutaneous approach, hence to prevent it, the adhesions have been created between the parietal pleura and GPB by modified Brompton Technique and then the air is suctioned out of bullae followed by intracavitary talc (sclerosing agent) insufflations to obliterate the air space by fibrosis.[10,11] The spirometry, 6 Minute Walk Test (6MWT) and imaging are the reproducible modalities helps in monitoring and assessing the prognosis of an individual. Sungrock Park *et al.* mentioned spontaneous regression of GPB which may be due the reabsorption of air after the blockage of draining airways either by inflammation, mucous plugs, infection or compression with its own pressure.[12] He further stated that at present there is no authentic guideline available for the management GPB, but somehow surgical resection is anonymously recommended. However percutaneous intra-cavitary drainage is the safest alternative in surgically unfit patients with comorbidities and or severely impaired cardiorespiratory function *(as present in our case).* We are presenting the preliminary management of a very rare surgically unfit case of GPB which was compressing the heart and thus declared unfit for surgery.

Case Presentation

A 27-year female was admitted with the chief complaints of shortness of breath (2 months) which has increased since the last 10 days, cough 10 days, fever off and on last 10 days and chest pain dull achy radiating to the back 10 days. Her general condition was poor with BP 90/60, Pulse 98, Respiratory rate 20 and SpO_2 90% on room air and the patient was admitted to the respiratory ICU. The O_2 at the rate of 2 to 3 lit/ minute with nasal prong was started. She was slightly pallor with an absence of cyanosis, Icterus, pedal edema and clubbing. Her JVP was raised. She had a past history of anti-TB Treatment for Tubercular lymphadenitis of neck. No history of DM/ HT/ Operation/ Blood transfusion. The investigations revealed Hb11.2 gm %, TLC 11200 and the rest of including blood sugar, serum protein, renal and liver profile, as well electrolytes were unremarkable. HIV/ HbsAg were non-reactive.

Her chest X-ray (Figure 1) revealed a single huge emphysematous bulla (GPB) almost occupying right hemi-thorax with the shift of the mediastinum to the opposite side. HRCT thorax (Figure 2A, 2B) further showed tension bulla occupying the azygos-esophageal recess with shift and mass-like compression of the heart and mediastinum. ECG showed sinus tachycardia, with T wave inversion in lead V5-6. ABG on O_2 support revealed 7.33/ 62/ 96/ 32.3 (partially compensated respiratory acidosis) due to chronic poor lung functions with reduced alveolar/ minute ventilation. Antibiotics and diuretics were given

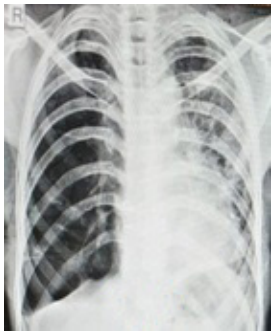
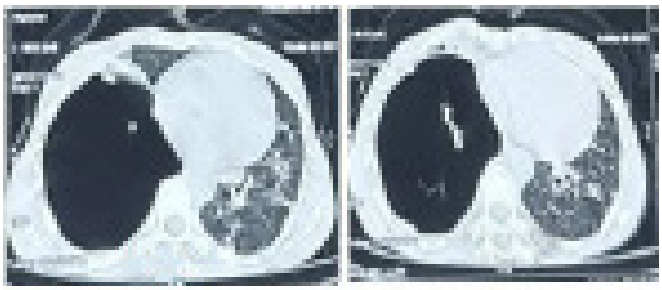


Figure 1: Chest X-ray



(A) **(B)**

Figure 2: HRCT thorax

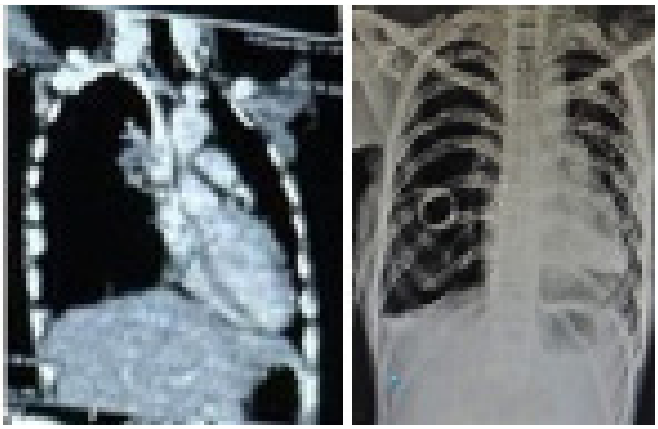


Figure 3: CXR

as part of the treatment. The culture and sensitivity report of sputum stated the growth of commensal flora after 24 hrs of aerobic incubation and no fungal element seen. ZN stain for AFB was negative. USG Neck reported old healed cervical TB Adenitis and USG abdomen stated renal calculi 6 mm size in the left lower calyx.

Our patient was referred from the private hospital and about 4-5 days prior a spirometry reported FVC (16%), FEV1 (17%) and FEV1/FVC (109%) with remarkably poor lung functions, and a 2D ECHO reported by a cardiologist as LVEF 30% with normal LV size and severe LV dysfunction, mild MR, Mild AR and reduced LV compliance. The patient as well her attendants refused for bullectomy at the higher centre so after written consent we have decided to decompress the giant bulla by the trans-cutaneous placement of a pigtail catheter with an underwater seal. The procedure remained uneventful (CXR Figure 3) with progressive relief in shortness of breath & chest pain.

A repeat 2D ECHO showed remarkable improvement of cardiac ejection fraction from 30% to 58% and a normal pulmonary arterial pressure of 20 mm. The patient got clinical improvement and satisfied fit for video-assisted thoracoscopy (VATS) and or bullectomy. The patient was discharged with the catheter in situ, to a higher center for surgical intervention.

Discussion

Our case was a rare presentation of idiopathic GPB. She was a young female without underlying preexisting lung disease especially the commonest etiology i.e. COPD and or smoking-related diseases. The unilateral lung (Right) was affected that too with single giant bullae, occupying most of the hemithorax and compressing rest of the healthy lung. The contralateral lung was also healthy. This was resembling with a group I, as mentioned by Avinash Aujayebet *et al*.[6] The mass-like effect on mediastinum with sign and symptoms along with markedly compromised cardiac dysfunction, documented by diminished ejection fraction on 2 D ECHO, all together were very rare, to make this case worth presenting. This patient got tremendous relief after a simple percutaneous placement of a pigtail catheter intrabullous with an underwater seal. The whole procedure was carried out under local anesthesia with markedly improved cardiac activity so it's another factor of interest. Bullectomy or surgical resection is gold standard in management but as and when with change in clinical scenario and considering the number of bullae, its size, site, and location (unilateral or bilateral) with the underlying condition of lung or an alternate minimal invasive procedure i.e. transcutaneous drainage with the underwater seal have opted.

Conclusion

The GPB may leads to chronic hypoxia and a mass-like effect so to avoid life-threatening emergency situations, and in surgically unfit cases, one has to select the result-oriented, minimally invasive procedure to decompress of GPB after considering risk and benefit.

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