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Vanishing Lung

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Abstract

Background: Vanishing lung is a rare syndrome. It mainly affects young males who are smokers, it considered an advanced stage of bullous disease, where the entire lobe or lung paranchym is replaced by bullae, it appears radiologically as a hyperlucency due to air trapping and destruction of interstitial tissue and vascularity in alveolar wall's. Misdiagnosed usually as pneumothorax so must be differentiated from other causes of Hyperlucency lung syndrom. Hereby a case of vanishing lung diagnosed primarily as a post TB lung destruction. **Case Report:** A sixteenth-year-old virgin female patient, with treated for TB for six months without radilogical improvement. CXR and CT scan revealed diffuse left lung hyperlucency, TB work up (sputum exam, washing by bronchoscopy) appears no active disease. Left pneumenctomy had done, grossly there are no lung pranchyma and microscopically no signs of TB in the specimen. The findings are consistent with Vanishing lung. **Conclusions:** An understanding of the broad differential diagnosis of pulmonary hyperlucency is necessary to determine the underlying cause and provide appropriate patient care.

Keywords

Vanishing Lung, Bullae, Post TB Lung Destruction, Lung Hyperlucency

1. Background

Hyperlucency is a common finding in CXR and chest CT scan, it means decreased density of chest component including chest wall or lung, appears due to technical problems (position asymmetry) or anatomical lesions, bilateral or unilateral, diffused or localized, and congenital or acquired can be caused by absence of the chest wall muscles and ribs, or by increased air trapping in the lung or intrathorax cavity, or decreased the lung paranchymal structures including interstitial tissues and vascularity, hereby in Figure 1, the most causes of this

finding [1].

2. Case Report

On October 2017, sixteenth year old virgin female patient, referred to our hospital after six month of TB treatment without radiological improvement.

Pt had breathlessness and recurrent lower respiratory tract infections since many years without any previous documents.

No history of positive smoking, no familial history of respiratory illness.

She was healthy, no cashecsia, no fever, no night sweating.

Left chest revealed decreased air entry on auscultation, and increased resonance on percusion.

Cxr: Diffuse hyperlucency of left hemithorax (Figure 2).

Chest CT scan: Absence of left lung paranchyma structure, and bullae in right lung apex (Figure 3).

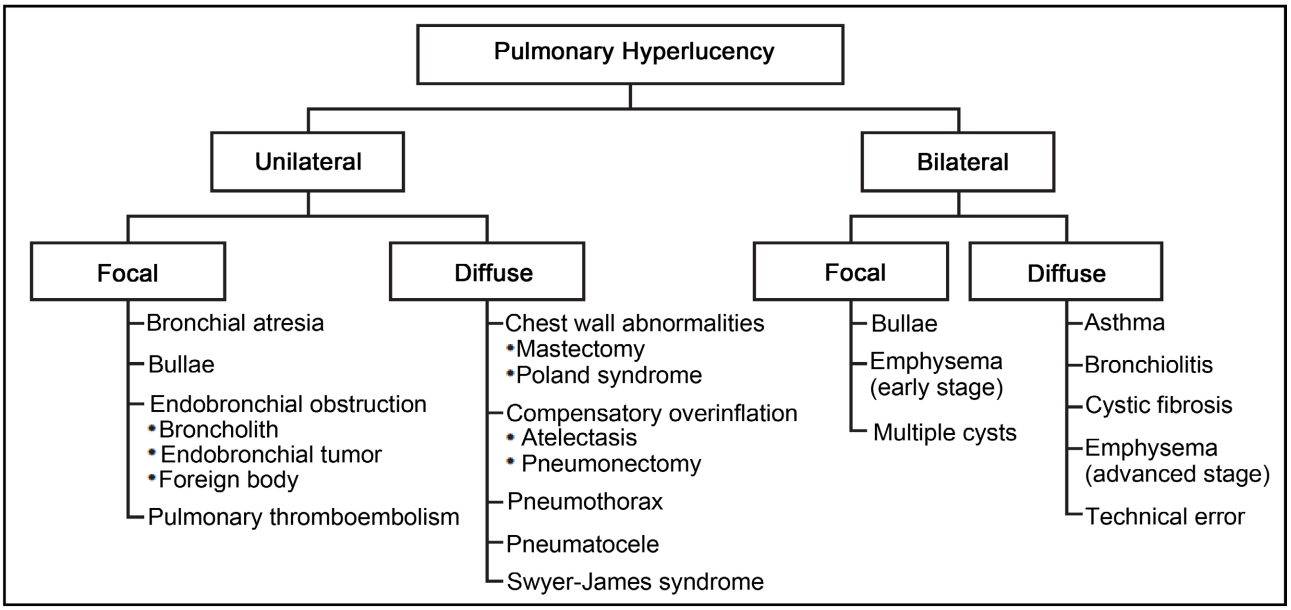

Figure 1. Flowchart shows differential diagnosis of unilateral and bilateral pulmonary hyperlucencies.

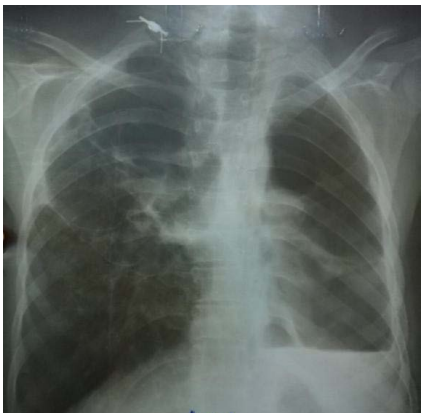

Figure 2. CXR: Diffuse hyperlucency of left lung.

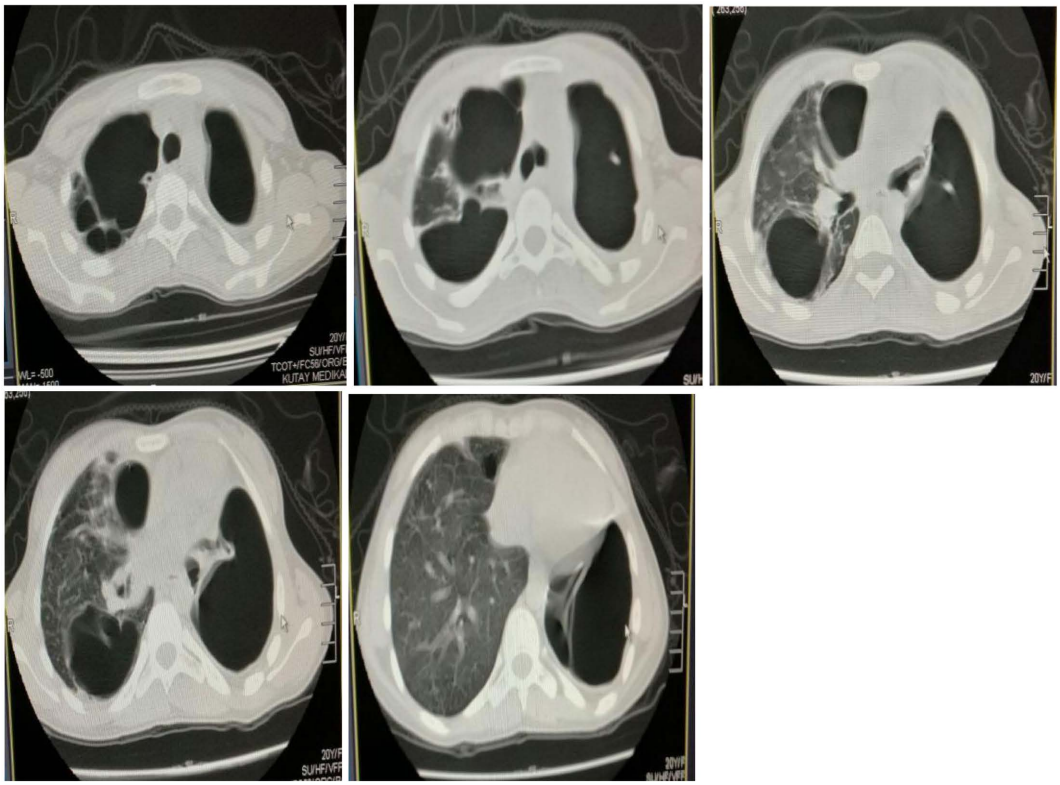

Figure 3. Chest CT scan: Absence of left lung paranchyma structure, and bullae in right lung apex.

Sputum was negative for AFB, koch bacilli, by ziehl-neelsen staining.

Bronchoscopy revealed normal broncheal tree with negative AFB, koch bacilli, in washing.

So the primary diagnosis was post TB left lung destruction, and left pneumonectomy has done. In the specimen the lung consist of many cavities without normal paranchyma (**Figure 4**).

Pt went well without any complications and discharged from the hospital within one week.

Pathology report (**Figure 5**) revealed:

- Left Lung: Presence of cavity composed of fibrous connective tissue, with signs of chronic non-specific inflammation.
- Pleural: Chronic non-specific fibrosing Pleuritis.
- No evidence of Tuberculous inflammation in the received specimen.
- No malignancy in the received specimen.

That consistent with the diagnosis of Vanishing lung.

On December 2021, she visited our clinic with lower respiratory infection.

She revealed that she went very well through the last five years with a mild respiratory infections during winter, and has married and had two kids.

During the follow up, CXR: small white left hemithorax and compensatory inflation of right lung (**Figure 6**).

Ct scan: resected left lung, left shifting of heart and mediastinum, compensatory inflation of right lung with same bullae in the apex (**Figure 7**).

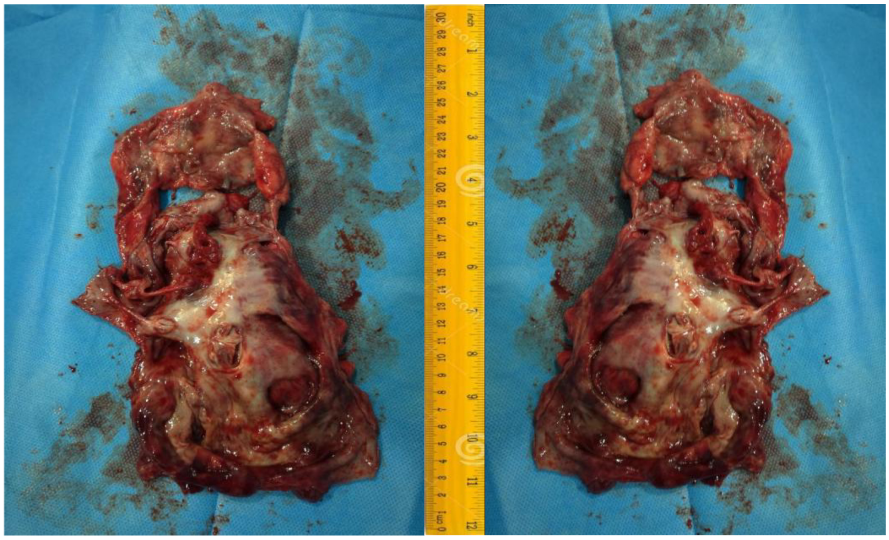

Figure 4. The resected lung.

Patient's name: F. M. J. Age: 16 Ys
Referred by: Date Received: 4 NOV
Specimen: Left Lung
Number: 11 - 18 / 17

PATHOLOGY REPORT

DESCRIPTION OF THE SPECIMEN:

Received in formalin is the left Lunge measures 14 X 7 cm, sectioning reveals cavity measures 6 cm in diameter, encountered by hemorrhagic pulmonary tissue, elastic firm in consistency, with presence of membranous pieces grayish in color, and elastic-firm in consistency.

MICROSCOPIC EXAMINATION:

Sections from the lung shows cavitary formation devoid of lining cells, composed by vascular fibrous connective tissue with zones of collections of chronic inflammatory cells.

The pleural sections show dense fibrous connective tissue moderately infiltrated with chronic inflammatory cells.

CONCLUSION;

- Left Lung: Presence of cavity composed of fibrous connective tissue, with signs of chronic non-specific inflammation.
- Pleural biopsy: Chronic non-specific fibrosing Pleuritis.
- No evidence of Tuberculous inflammation in the received specimen.
- No malignancy in the received specimen.
- No malignancy.

6 NOV 2017

Dr. Fadi SAKKAL

Figure 5. Pathology report.

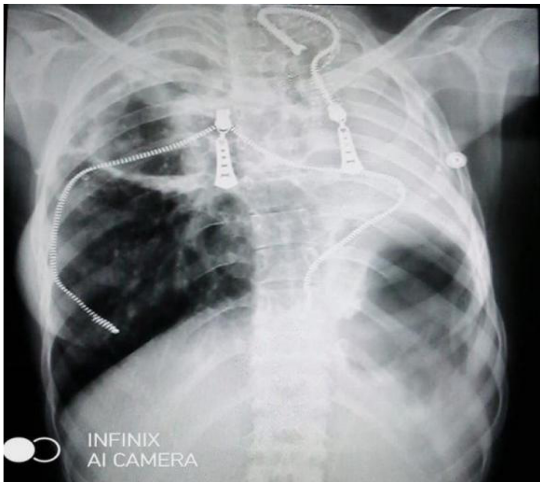

Figure 6. CXR: small white left hemithorax and compensatory inflation of right lung.

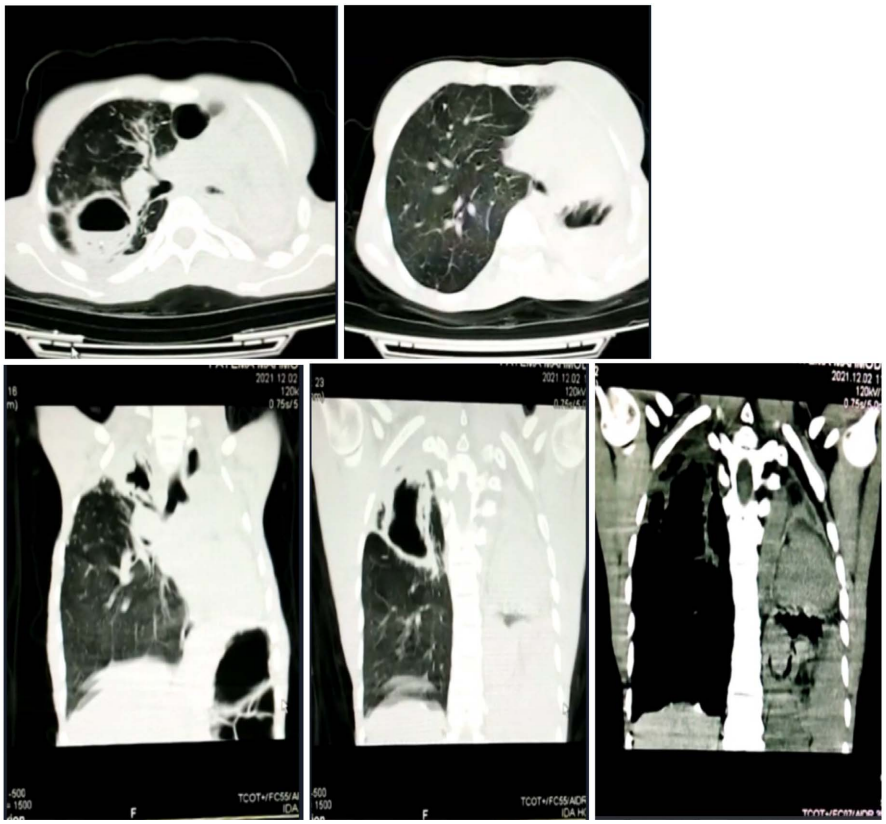

Figure 7. CT scan: resected left lung, left shifting of heart and mediastinum, compensatory inflation of right lung with same bullae in the apex.

3. Discussion

The diagnosis of our case has been confused between lung destruction after TB and vanishing lung, since in both lesions, bullae and cavities can be found.

In Vanishing lung syndrome the entire lobe or lung normal lung paranchym is disappeared and replaced by *bullae*.

Bulla are air-filled spaces within the parenchyma that are 1 cm or larger in diameter, less 1 cm called bleb, and consist of a thin wall of visceral pleura with remnants of alveolar and interlobular septa inside [2].

If bulla occupy more than 30% of the chest cavity is called a *giant bulla*. Bulla origins within a normal lung tissue in 20% and within emphysematous lung in 80%, *Bullous emphysema*. Several classifications for giant bulla depending on the condition of the underlying lung paranchyma established (**Table 1** and **Table 2**) [3].

Giant bullae is usually unilateral and asymmetric and affecting young patients; while, bullous emphysema has bilateral involvement and affecting elderly patients. The major cause of bullae formation is cigarette Smoking [2]. Indications of surgery in bullous disease include dyspnea and bulla complications, the complications include: pneumothorax, bulla infection, concurrent lung cancer and hemoptysis. The best results can be achieved in patients with gaint bulla compressing normal lung paranchyma and low progress of dyspenea. The goal of surgery is to resect the bulla by stapller and let the underlying collapsed normal lung to re-inflate, that can be done by thorachotomy or thoracoscopy or median sternotomy. Unless an operation for cancer is being performed, anatomic resections are generally avoided. Even with apparently largely destroyed lung, there is often functional tissue near the hilum. Rarely, an entire lobe is destroyed and an anatomic resection is appropriate [3]. Relative contraindications for these operations

Table 1. DeVries and wolfe classifi cation of giant bullae*.

Group	Bullae	Underlying Lung
I	Single large	Normal
II	Multiple	Normal
III	Multiple	Diffuse emphysema
IV	Multiple	Other lung diseases

*Based on the nature of the bullae and the underlying lung. From DeVries WC, Wolfe WG: The management of spontaneous pneumothorax and bullous emphysema. Surg Clin North Am 60:8, 1980.

Table 2. Witz and roeslin classifi cation of giant bullae*.

Group	
I	Bullae with normal underlying parenchyma, paraseptal emphysema
II	Bullae with diffuse emphysema; bullae are a local exacerbation of diffuse panacinar emphysema
III	Vanishing lung syndrome; entire lobe or lung replaced by bullae

*Based on underlying lung. From Witz JP, Roeslin N: La chirurgie de l'emphyseme bulleux chez l'adulte. Revue Francais de Maladies Respiratoire 8:121, 1980; as in Mehran RJ, Deslauriers J: Indications for surgery and patient work-up for bullectomy. Chest Surg Clin North Am 5: 717-734, 1995.

include hypoxemia, hypercapnia and COPD.

All diffuse hyperlucency of lung coming as a differential diagnosis of vanishing lung. TB, which is endemic disease in some reigns in our country, considered a greater imitator of this case.

Endobronchial TB may result in bronchial stenosis. The damage may lead to secretion retention and chronic infection leading to bronchiectasis or abscess formation. Partial obstruction may lead to lobar collapse or the development of bullae and hyperinflation (**Figure 8**). As often the distal lung is damaged beyond salvage, resection is often indicated [4].

Bronchiectasis (**Figure 9**) may occur as a direct complication of tuberculosis

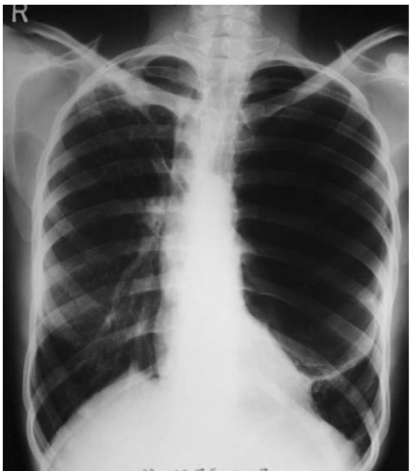

Figure 8. Bulla secondary to tuberculosis. Middle-aged patient with known past history of treated tuberculosis presented with increasing shortness of breath. Chest radiograph shows a large bullae in the left hemithorax. Bronchoscopy showed a stenosis of the left upper lobe bronchial orifice. Treated with a left upper lobectomy.

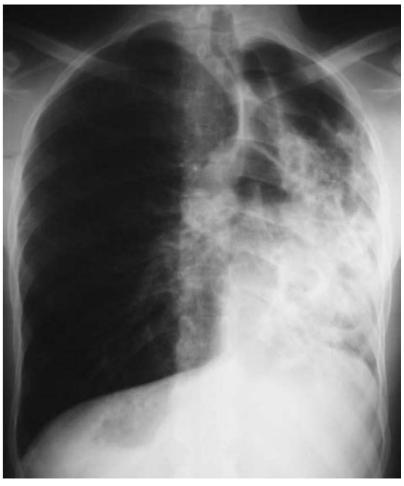

Figure 9. Total lung bronchiectasis secondary to tuberculosis. Young male patient presented with recurrent lung infections. Treated for tuberculosis on two prior occasions. Chest radiograph shows a completely destroyed left lung. Treated with a left pneumonectomy.

infection or as a result of superimposed, often multiple, bacterial infections. The pathology of bronchial dilation, chronic pooling of secretions leads to chronic fetid sputum and recurrent local and distant sepsis formation with the risk of serious complications such as hemoptysis or metastatic abscess formation.

The treatment is generally medical for minimally symptomatic disease or for instances where it is diffuse or multifocal. Where complications are evident and the disease localized, resection may be the treatment of choice [4].

Vanishing lung also usually can be confused with pnumothorax or tension pneumothorax and may be mismanaged by chest tube, Ct scan playing a big role in distinguishing between the two cases depending on the double wall signs [5] [6].

4. Conclusion

An understanding of the broad differential diagnosis of pulmonary hyperlucency is necessary to determine the underlying cause and provide appropriate patient care [1]. Regarding bullous disease, in properly selected patients, most can be expected to have subjective improvement of their dyspnea as well as demonstrable improvement in pulmonary function testing. The long-term of this improvement is dependent on the quality of the underlying lung and the progression of any disease in that lung [3].

Consent

A written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Conflicts of Interest

The author declares that he has no competing interests.

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