



Case report

Surgical treatment of giant bullae on the background of cystic lesion and vascular malformation. Case report

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ABSTRACT

Bullae formation on a cystic background is still not described thoroughly. This clinical case aims to improve this situation and describes the combination of multiple cysts with vascular malformation, giant bullae (2/3 of the left hemithorax and 1/2 of the right hemithorax) with suppuration in a 50-year-old smoker. Before surgery, treatment for chronic obstructive lung disease (COPD) was administered without a positive outcome. Bullectomy was subsequently conducted and histopathological data revealed vascular malformation causing micro-infarctions in pulmonary parenchyma, cystic degeneration and tuberculosis. Although it became possible to establish a final diagnosis due to histology, the need for surgical treatment was clear from clinical and x-ray evidence.

Such instances as the one presented in this case report should assist with improving our knowledge about bullae in combination with cysts in the lungs because they provide additional context in a clinical setting.

1. Introduction

Cysts are epithelial-lined cavities and often are congenital structures [1–3]. Therefore, the presence of cysts does not preclude the formation in lung cavities of other etiology. The formation of bullae within a background of cystic lung changes is not well understood. Smoking is known to be a factor affecting the lung's elastic framework and a risk factor for bullous changes. On the other hand, little information exists regarding any link between the formation of cysts and bullae. Different complications of bullae disease may develop, in which there are indications for surgical intervention: a tense bullous cyst and fluid accumulation in bullous cysts. These complications are much less common than pneumothorax. Therefore, they are of interest for clinicians as to how to diagnose and treat those conditions due to the present dearth of clinical information about this. The combination of polycystic with vascular malformations, giant bullae with suppuration [4,5] and tuberculosis is additionally of significance for clinicians, when integrated with an understanding of the pathogenesis of vanishing lung syndrome and a patient's prognosis. Unfortunately, we have found only one previous published clinical case of combined cysts and giant bullae [2]. This paper attempts to partially rectify this by presenting another

such clinical case as presented below.

2. Clinical case

A 50-year-old male, smoker (50 packs/year), was diagnosed in 2000 with chronic obstructive lung disease (COPD) and was admitted to a local hospital complaining of chest pain, cough with purulent sputum up to 150 ml/day, dyspnea on exertion and fever up to 39°C. He was treated with Amoxicillin, Tiotropium bromide and Salmeterol.

Due to the failure of conservative treatment, the patient was routed to the thoracic surgical clinic. A chest X-ray and CT scan revealed giant bullae in the upper parts of the lungs (2/3 of the left hemithorax and 1/2 of the right hemithorax) [6]. Some bullae in the right lung contained fluid. The lung tissue was emphysematous in the epiphrenic regions, but did not contain bullae (Fig. 1).

During further examination, tuberculin tests were negative; acid fast bacilli (AFB) were not detected in the sputum neither microscopically nor by culturing.

Spirometric parameters, the partial pressures of blood gases and 6-min walking test were decreased compared to normal (Table 1).

Taking into account the spirometry indicators' discrepancy to COPD GOLD criteria, the absence of alpha1 antitrypsin level decreasing, we

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Abbreviations

COPD	chronic obstructive lung disease
FEV1	forced expiratory volume in 1 second
FVC	forced vital capacity
VATS	video-assisted thoracic surgery
VC	vital capacity

initially diagnosed bullous lung disease (vanishing lung syndrome) [7] complicated by non-specific suppuration in the right lung with sputum culture revealed *Streptococcus viridans* at 3×10^7 . Due to chronic infection persistence, not amenable to conservative therapy, and progressive respiratory failure, then a surgical intervention was the only option for this patient [8].

At the first stage, the patient underwent a thoracoscopic lobectomy of the right upper lobe en-bloc with segment 6.

At the second stage, the patient underwent a trisegmentectomy of the left upper lobe six weeks later. (Fig. 2).

Histological examination (Fig. 3) revealed:

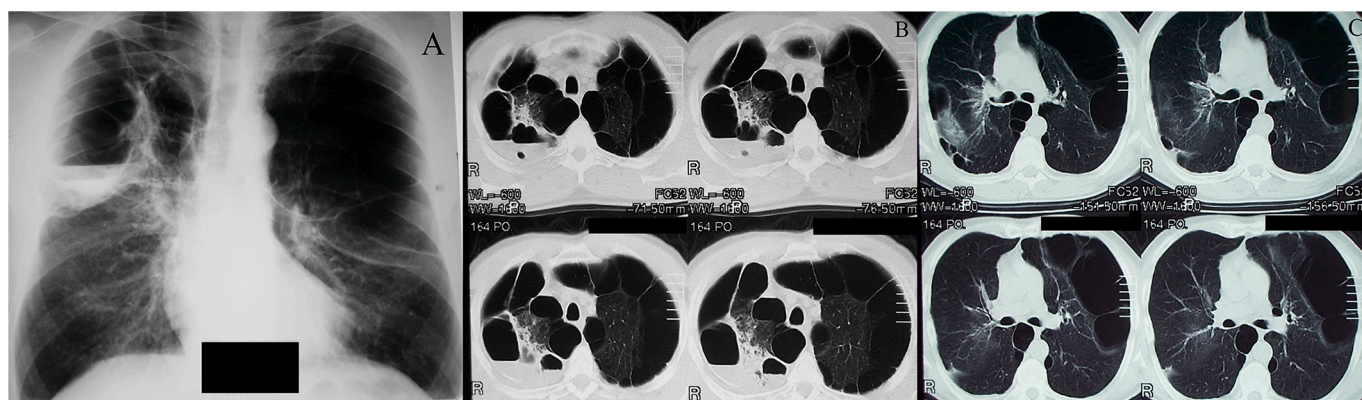


Fig. 1. Roentgenological examination.

A - Chest X-ray on admission. Bullae and giant bullae are evident in both lungs. With the fluid is presented in the right lung.

B - Chest CT on admission. Giant bullae occupies upper parts of both lung. In the upper lobe of the right lung there are some cavities containing fluid.

C - Chest CT on admission. In the middle sections, the lung tissue is preserved, emphysema is less pronounced, single medium-sized “bullae” on the right and the continuation of the system of giant bullae on the left are visible.

Table 1

Patients’ lung function characteristics.

Parameters	Before surgery	3 month after surgery	10 years after surgery.
Blood analysis	leucocytosis - $14,3 \cdot 10^3/L$ ESR 38 mm/h	leucocytosis - $9,5 \cdot 10^3/L$ ESR 17 mm/h.	No data
6 minute walk test (normal for a healthy men is approximately 550–600m [12])	432m	428m	620m
Spyrometry	VC 67%, FEV1 63%, negative	VC 64%, FEV1 61%, negative	VC 92%, FEV1 78%, negative
Test with bronchodilator (Fenoterol 400 mcg)	negative	negative	negative
Blood gases	pCO2 - 40,6 mm Hg pO2 - 65 mm Hg	pCO2 - 38,7 mm Hg pO2 - 74 mm Hg	pCO2 - mm Hg pO2 - mm Hg
MEF 50		58%	50%

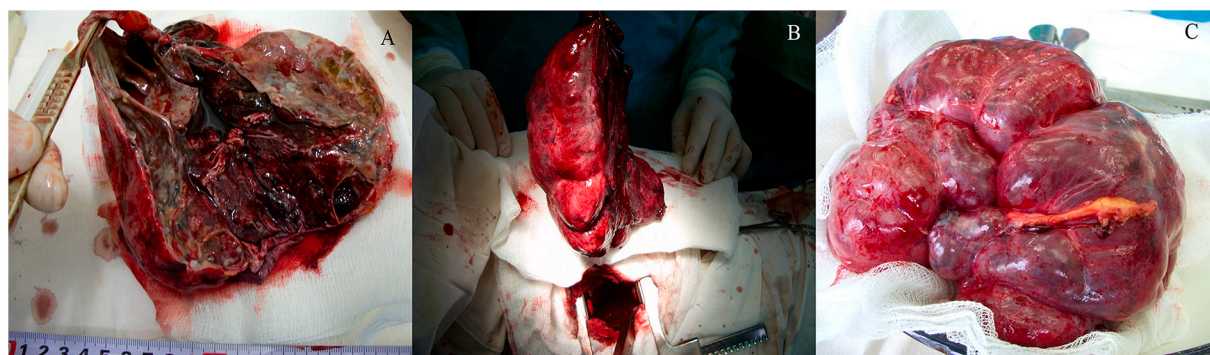


Fig. 2. Cross -section of the removed parts of the right lung.

A - The lobe contains multiple cavities with purulent contents.

B - Stage of the second operation: removal of the lung - I, II, III segments of the left lung upper lobe.

C - Removed lung in the second operation. I, II, III segments of the left lung upper lobe.

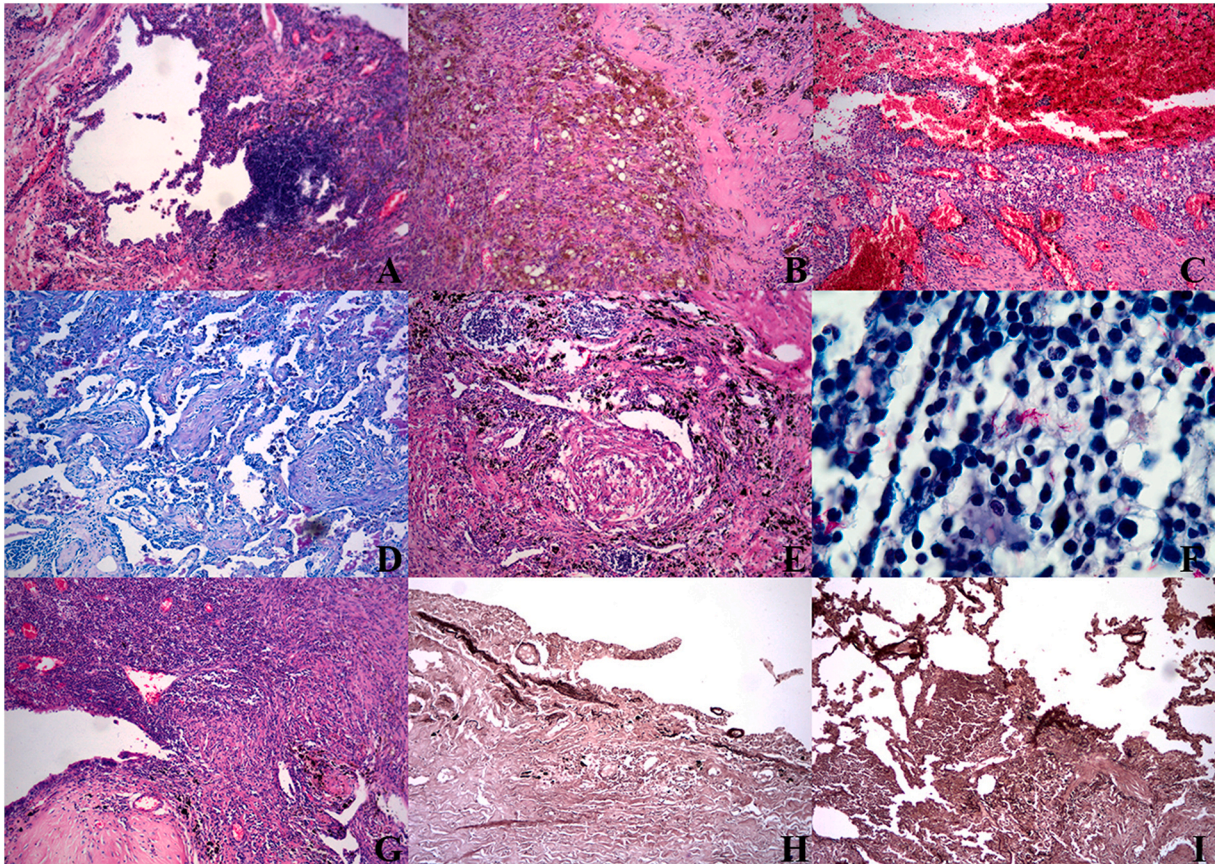


Fig. 3. Histological samples.

- A - forming bulla of lung tissue, next to cysts in a common zone of exacerbation of chronic inflammation and the presence of an adjacent vascular malformation, magnification $\times 40$, stained with hematoxylin and eosin);
- B - area of organizing hemorrhagic infarction next to lung tissue cysts and vascular malformation, presence of foci of nonspecific inflammation, magnification $\times 100$, stained with hematoxylin and eosin;
- C - Fresh hemorrhage near vascular malformation with perifocal inflammatory reaction, magnification $\times 100$, stained with hematoxylin and eosin;
- D - Next to the infarction area, an area of organizing pneumonia, magnification $\times 100$, Mallory stain;
- E - Fibrotic tuberculous granuloma in a zone of a fibrocystic modified lung, on the periphery of an organizing hemorrhagic infarction in the lung tissue. Visible accumulations of hemosiderophages. The granuloma is characterized by indiscriminate fibrosis, the presence of a giant cell reaction and a small number of leukocytes. On the periphery, an epithelioid cell reaction with an admixture of leukocytes remains and accumulations of lymphocytes are formed, magnification $\times 100$, stained with hematoxylin and eosin;
- F - Acid resistant mycobacterium tuberculosis in the area of necrosis with severe tuberculous inflammation, magnification $\times 1000$, Ziehl-Neelsen stain);
- G - Section of the wall of cystic dilated bronchus with a fibro-altered wall after healing of the focus of tuberculous inflammation, a round section of a slightly vascular coarse fibrous fibrosis, with significant fibrous changes in the adjacent lung tissue having the presence of epithelioid cell infiltration with an admixture of leukocytes and an area of adjacent vascular malformation, magnification $\times 100$, stained with hematoxylin and eosin;
- H - Fragmentation and disappearance of elastic fibers in the zone of formed cysts and fibro-altered lung tissue, magnification $\times 100$, stained with acidic orcein;
- I - The disappearance of the elastic framework in the interalveolar septa and its preservation only in the vascular walls during the formation of bullous changes in the lung tissue in the area of inflammation, magnification $\times 100$, stained with acidic orcein.



Fig. 4. At the control examination of the patient, 10 years after surgical treatment: A - a survey radiograph of the lungs; B - a photography of the patient.

- bullae of lung tissue with multiple cysts in the area of chronic inflammation with the presence of vascular malformation
- zones of organizing hemorrhagic infarcts next to lung cysts and vascular malformations,
- foci of non-specific inflammation
- fibrotic tuberculosis granuloma in the area of fibrocystic modified lung
- cystic change of the bronchi.

The histological report established the diagnosis: bullous disease type I (vanishing lung syndrome) complicated by suppuration (*Streptococcus viridans*). Polycystic lung with vascular malformation of the lung parenchyma. Small focal pulmonary tuberculosis [3].

Antibiotic therapy was carried out for 3 weeks in accordance with bacterial sensitivity data. On discharge, 3 months post-surgery, the patient's condition was satisfactory with the signs of respiratory failure having significantly decreased. At a follow-up examination, 10 years later, the patient's status condition was good. He has continued to smoke. A chest X-ray did not demonstrate new bullae (Fig. 4). Spirometric parameters were satisfactory (Table 1).

Positive dynamics of functional parameters and gas exchange was noted in comparison with previous data (Table 1).

3. Conclusions

In this clinical example, a background of cystic lung changes was presented. The vascular malformation role isn't clear there. Multiple bullae appeared and gave a picture of the vanishing lung (bullous lung disease I type), complicated by non-specific suppuration [2,8–11] and also combined with only histological signs of tuberculosis. Due to the absence of COPD in this patient, the presence of these conditions was determined only on the basis of histopathological data. The question is still open as to why there were not immunological, clinical and roentgenological signs. Although it became possible to establish a final diagnosis due to histology, the need for surgical treatment was clear from clinical and x-ray evidence. This case report provided us with better knowledge about bullae in combination with cysts. Additional published case studies would be therefore a welcome addition to the literature.

Author statement

Giller D.B.: Conceptualization, Validation, Writing - Review &

Editing, Supervision.

Severova L.P.: Methodology, Writing - Review & Editing, Visualization.

Giller B.D.: Investigation, Writing - Review & Editing, Visualization.

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Martel I.I.: Methodology, Validation, Investigation, Writing - Original Draft.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.rmcr.2020.101198>.

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