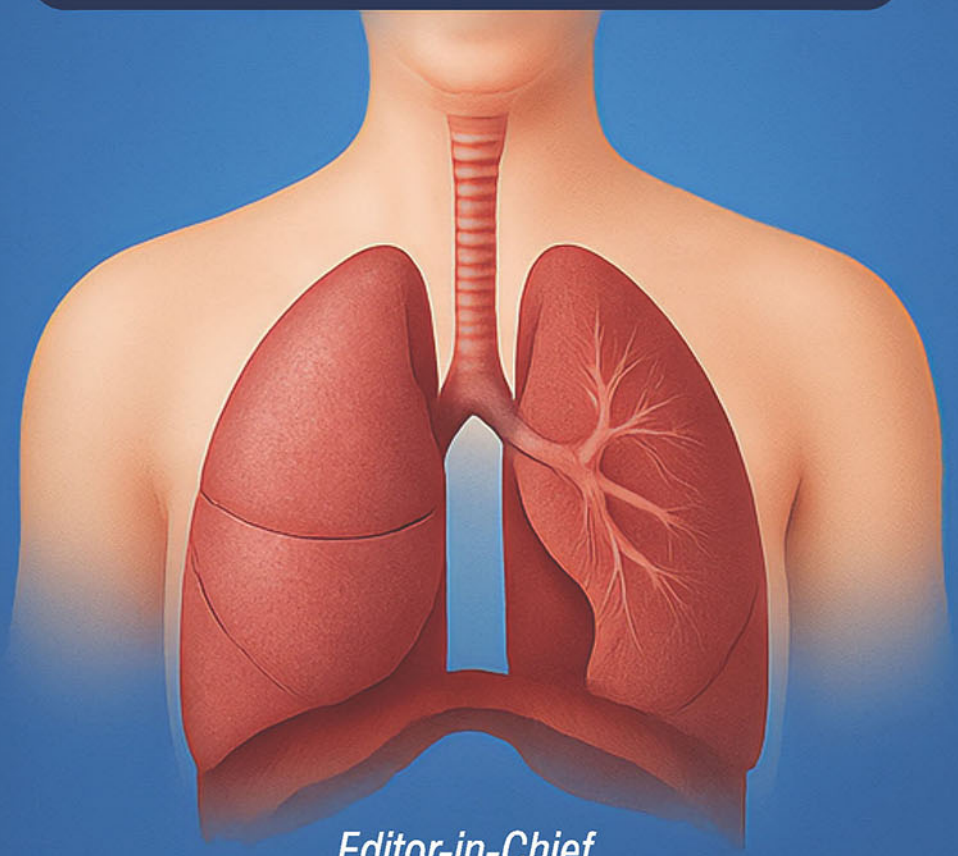
# Breathe Easy Pearls and Pitfalls in Respiratory Medicine

## A Compendium of Clinical Cases



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# CASE 10

# Persistent Right-sided Pleural Effusion: A Rare Etiology


**ABSTRACT**

A middle-aged woman presented with recurrent right-sided exudative pleural effusion. Initial evaluations were inconclusive, leading to empirical anti-tubercular therapy. Further intervention with thoracotomy revealed an esophageal duplication cyst (EDC) within the pleural cavity, causing persistent effusion. This case underscores the importance of considering rare congenital anomalies in the differential diagnosis of pleural effusions.


## INTRODUCTION

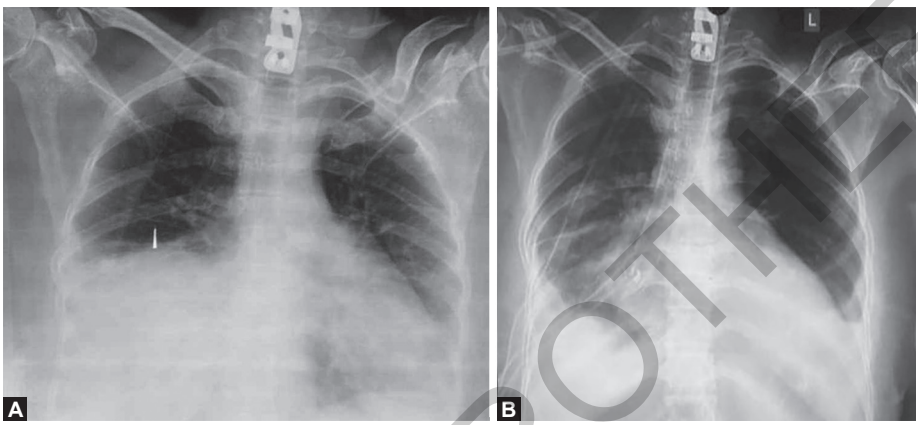
Esophageal duplication cysts (EDCs) are rare congenital malformations of the foregut, often presenting in childhood but might manifest in adults too. These cysts can cause compressive symptoms and are challenging to diagnose due to their rarity and the similarity in appearance to other mediastinal masses. This case report discusses a middle-aged lady with an EDC causing persistent right-sided pleural effusion.

## CASE PRESENTATION

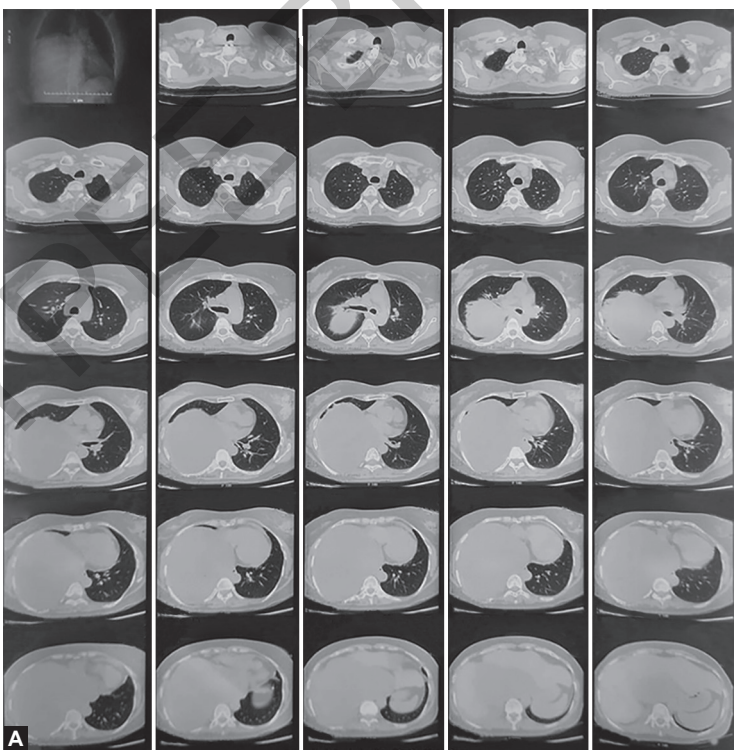
- *Patient information*: Middle-aged woman with a history of right-sided recurrent exudative pleural fluid effusion **(Figs. 1A and B)**
- *Initial investigations*:
  - *Pleural fluid analysis*: Lymphocyte predominant, negative for both malignancy, and cartridge-based nucleic acid amplification test (CB-NAAT) for TB
  - *Empirical treatment*: Anti-tubercular therapy initiated
  - *Repeat fluid analysis:*
    - Ordinary culture, fungal mount, acid-fast bacillus (AFB) stain, GeneXpert, and cytology negative.
    - *Fluid amylase*: 358 U/L, lipase: 5.6 U/L, carcinoembryonic antigen (CEA): 16,659 ng/mL, and adenosine deaminase (ADA): 99.4 U/L
- *Interventions*:
  - *Chest tube drainage*: Initial chest tube followed by a pig tail drain due to accidental removal of the chest tube.

- *Drain output*: Consistently around 50–100 mL of dark coffee-black-colored fluid

- *Advanced imaging*:
  - *Contrast-enhanced computed tomography (CECT) chest and abdomen*: Right-sided effusion with the possibility of mediastinal cysts and intra-abdominal communication **(Figs. 2A to C)**.




**FIGS. 1A AND B:** X-ray chest showing right-sided pleural effusion.




**FIGS. 2A TO C:** *Continued*

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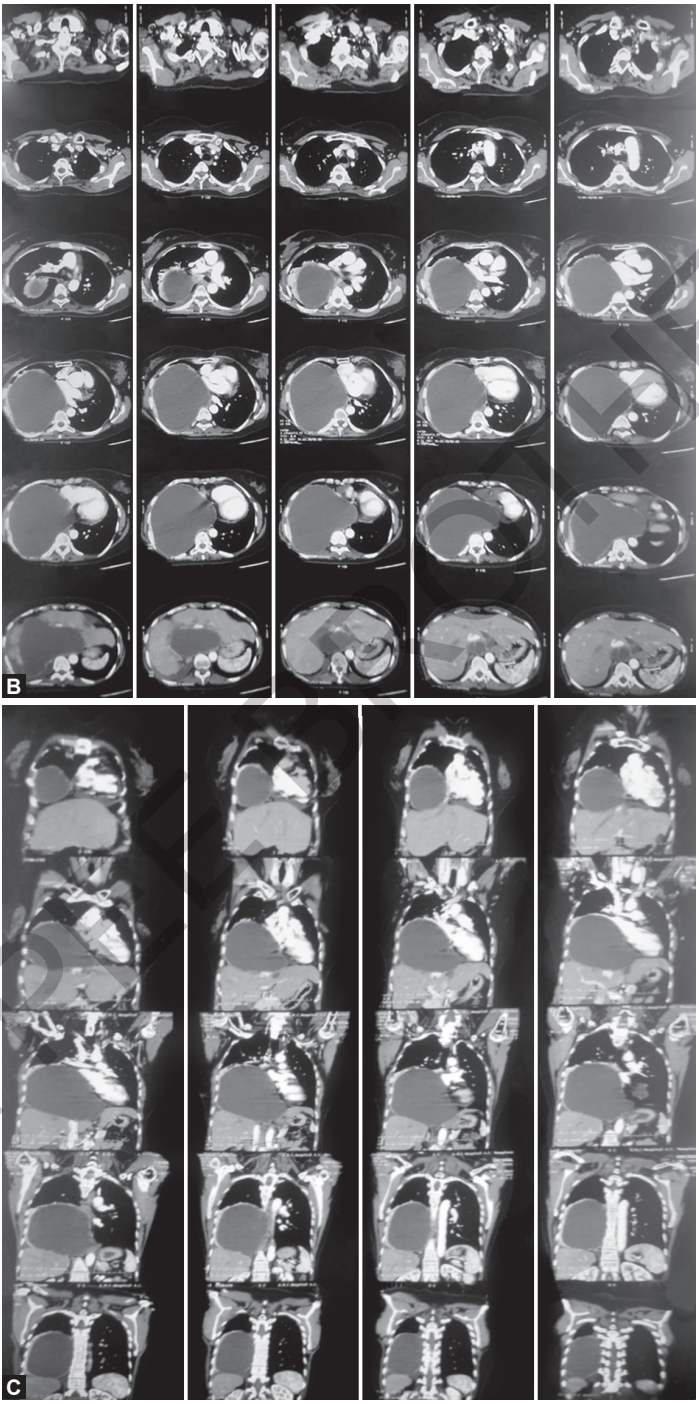
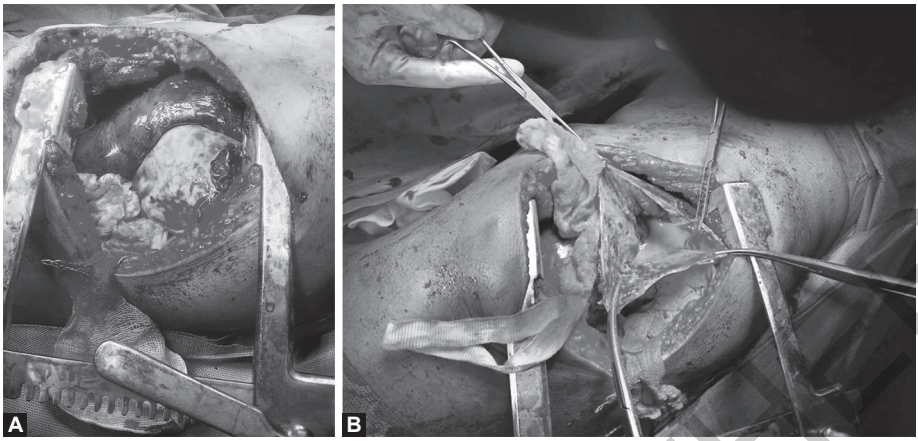



**FIGS. 2A TO C:** Contrast-enhanced computed tomography (CECT) chest showing right-sided effusion and mediastinal cyst.




**FIGS. 3A AND B:** Operative image showing opened pleura with the esophageal duplication cyst and residual fluid. ***(For color version, see Plate 1)***

- *Magnetic resonance cholangiopancreatography (MRCP)*: Showed cholelithiasis, loculated collection into the right mediastinum and abdomen, indenting the left lobe of the liver, inferior vena cava, and left hepatic vein.
- *Positron emission tomography (PET) scan*: Done to rule out any occult primary; however, showed a lesion in the right hemithorax without increased F-18 fluorodeoxyglucose (FDG) uptake

- *Surgical consultation*: Cardiothoracic and vascular surgery (CTVS) opinion sought; patient taken up for exploratory thoracotomy **(Figs. 3A and B)**.
- *Histopathology*: Cyst wall lined by pseudostratified columnar epithelium over a double smooth muscle wall **(Figs. 4A to D)**.
- *Final diagnosis*: Esophageal duplication cyst within the pleural cavity

## DISCUSSION

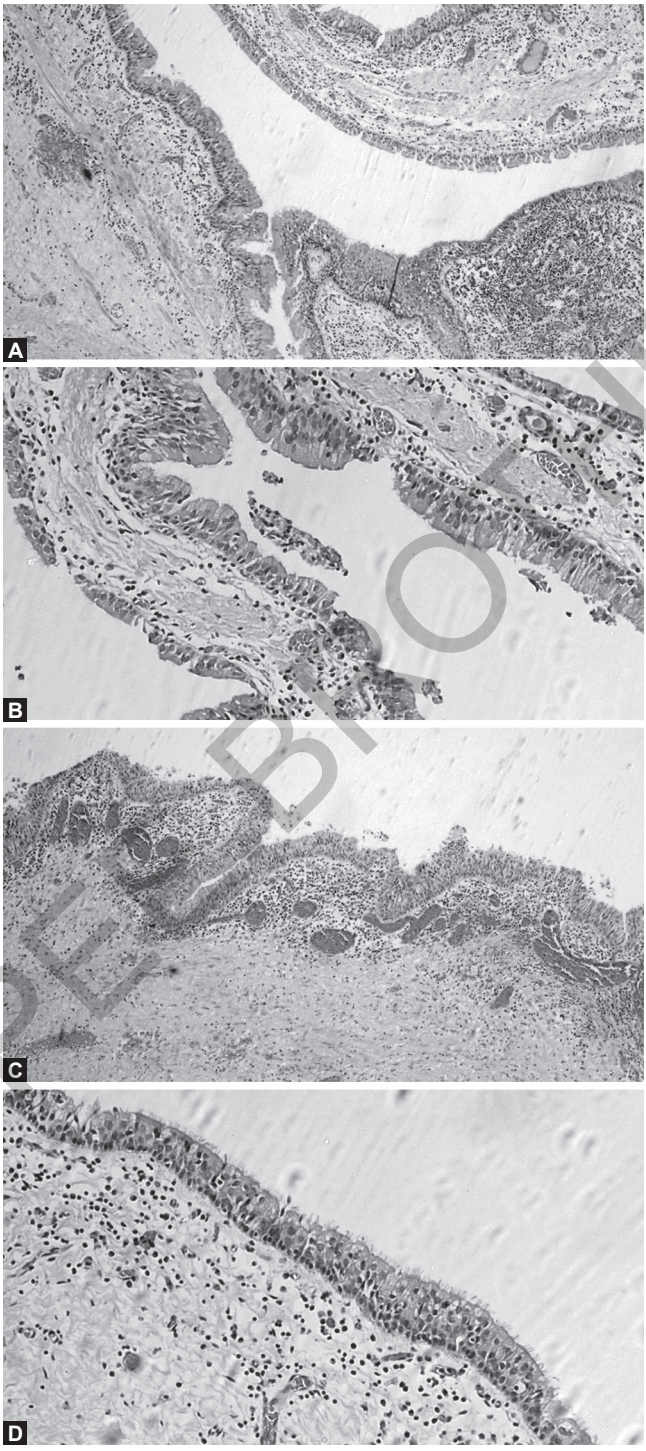
### Clinical Discussion

Esophageal duplication cysts are rare congenital anomalies, representing approximately 10% of all mediastinal tumors in children. They can present with varied symptoms, often depending on their size and location. An estimated 10–15% of all duplications cysts that occur in the alimentary tract stem from the esophagus.

In adults, EDCs can cause persistent and recurrent retrocardiac pleural effusions, as seen in this case. The high CEA levels in the pleural fluid were indicative of a malignant process, necessitating further evaluation and imaging. Only a handful of cases of EDCs within the pleural cavity and presenting with pleural effusions have been reported so far.

### Imaging Discussion

The initial chest X-ray and fluid analyses were inconclusive. Advanced imaging techniques like CECT and MRCP were crucial in identifying the cystic nature of




**FIGS. 4A TO D:** Histopathology of the cyst wall lined by pseudostratified columnar or mixed epithelial lining, overlying a double layer of smooth muscle. ***(For color version, see Plate 2)***

the lesion and its communication with the mediastinum and abdomen. PET scan findings of a lesion without increased FDG uptake further supported the diagnosis of a benign process, though malignancy could not be entirely ruled out without surgical exploration and histopathology. However, the exact nature and origin of the cyst are determined only by thoracotomy and subsequent histopathological analysis.

### Pathologic Discussion

Esophageal duplication cysts result from the aberrant development of the posterior division of the primitive foregut during embryogenesis. These cysts can occur anywhere along the esophagus, with 60% located in the lower third, which results in compressive dysphagia being the most common presenting symptom.[1,2]

Diagnosis can be challenging due to their rarity and the need to differentiate from other mediastinal masses. It has to be distinguished from many mediastinal masses, such as bronchogenic cysts, mature cystic teratomas, pericardial cysts, congenital cystic adenomatoid malformations, and neurogenic tumors. These mediastinal masses may have similar appearances as determined by ultrasonography, X-rays, and even CT visualization.

Thoracotomy and surgical exploration, along with histopathological examination, confirmed the diagnosis in this case.

## BRIEF REVIEW OF LITERATURE

Esophageal duplication cysts account for 10–15% of all duplication cysts in the alimentary tract and can mimic other mediastinal masses. Early diagnosis and appropriate surgical intervention are critical in preventing complications such as infection or rupture. This case emphasizes the need for thorough investigation and consideration of rare congenital anomalies in patients with persistent pleural effusions.

### *Clinical Pearls*

- EDCs should be considered in the differential diagnosis of unexplained recurrent pleural effusions.
- *Advanced imaging*: Techniques such as CECT, MRCP, and PET scans are crucial in identifying and characterizing mediastinal masses.
- Surgical exploration remains the definitive diagnostic and therapeutic approach for suspected EDCs.

## REFERENCES

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2. Cuch B, Nachulewicz P, Wieczorek AP, Wozniak M, Pac-Kozuchowska E. Esophageal Duplication Cyst Treated Thoracoscopically During the Neonatal Period: Clinical Case Report. Medicine (Baltimore). 2015;94(49):e2270.

# Breathe Easy: Pearls and Pitfalls in Respiratory Medicine

## *A Compendium of Clinical Cases*

In the present context, clinicians often come across a regular disconnect between the theoretical knowledge to application and interpretation of this knowledge in terms of patient care. This book is an effort to bridge the gap. The emphasis is on applied clinical medicine and problem solving which is not a part of regular postgraduate medical curriculum. In modern-day medicine, diagnostics, their importance, and interpretation have largely superseded clinical medicine. Through this varied case compilation, there is a conscious effort to holistically integrate and supplant investigational medicine with clinical medicine without undermining its importance.

***Highlights:***

- Comprehensive coverage of structured clinical cases, spotters, and rare respiratory cases
- Each chapter has learning objectives and ends with clinical pearls to reinforce these learning points
- Specific focus on management complexities and dilemmas
- Includes clinical correlation with radiological, pathological, and spirometry interpretations using a holistic approach for a better understanding
- The inclusion of flowcharts and references enhances educational value
- A handy clinical aid for pulmonologists, physicians, postgraduates, and clinicians alike
- Meets the curriculum requirements and can be used in university classrooms and seminars as a resource for case discussion and as a small group teaching exercise

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*Printed in India*

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ISBN 978-93-6616-758-9