

Pleuropericardial Cyst - A Rare Tumour of Mediastinum

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Abstract: *Pleuropericardial cysts (PPCs), account for 5 - 10% of all mediastinal tumors, are rare lesions occurring in approximately 1 in 100000 persons. Mostly they are detected postmortem or incidentally on routine chest X-ray (CXR). Multi detector Computer Tomography is used to confirm the diagnosis and are usually congenital and rarely acquired. Benign course and clinical latency are characteristic features of these cysts and the development of complications is rare, so that the majority of them can be left untreated(1).*

Keywords: Pleuropericardial cyst, mediastinal tumors, chest X-ray, congenital cyst, benign lesions

1. Introduction

Pleuropericardial cysts develop due to irregularities during the formation of the pericardial coelom in an embryo. The pericardial coelom later becomes the pericardial cavity, the space around the heart. If something goes wrong during its formation, it can lead to the development of these cysts. They are lined with a single layer of mesothelial cells and are filled with clear, watery fluid. The exact reasons for these developmental issues occur are not completely understood(2).

2. Case Report

60 year old male patient presented with fever on evaluation CECT chest shows COPD changes in bilateral lung fields predominantly the upper lobes. Well defined hypodense cystic lesion mea 7.5x6.6cm seen in the anterior mediastinum along the right side with post contrast peripheral enhancement. No evidence of septations or solid components within the cystic lesion. Possibly thymic cyst/pleuropericardial cyst. Cyst is abutting the SVC, ascending aorta medially, anteriorly the lesion abuts the chest wall, and medially it is causing subtle compressive atelectasis of adjacent parenchyma.

Patient underwent Video assisted thoracic surgery. We received a collapsed cyst and multiple grey-white membranous tissue. Cyst mea 5.5x5.5x3cm. Maximum wall thickness 0.2cm. Membranous tissue cut section grey- white myxoid. Microscopy shows partly cystic and solid tissue with mesothelial lining and extensive areas of chondromyxoid change. Immunohistochemistry shows EMA positivity in the mesothelial lining, S100 positive in the solid area, Ki 67 low proliferation. Lung tissue received shows extensive areas of haemorrhage and fibrosis. Final diagnosis was consistent with pleuropericardial mesothelial cyst with chondromyxoid change in the wall.

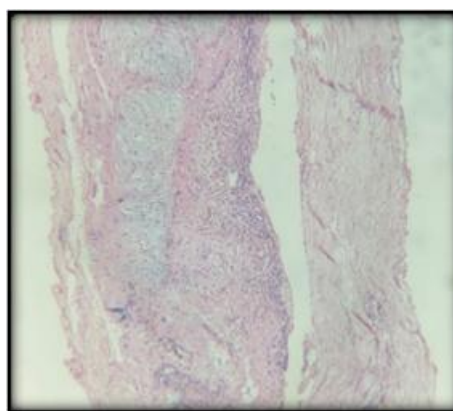


Figure 1

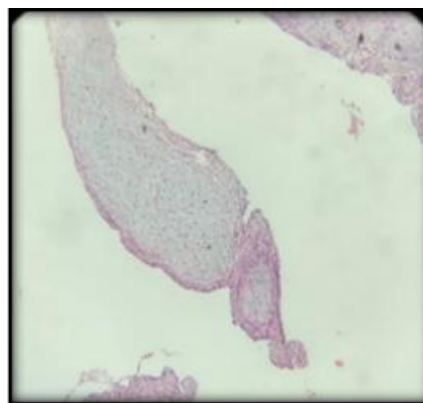


Figure 2

Figure 1, 2, 3, 4: Partially cystic and partially solid with mesothelial lining and extensive chondromyxoid change

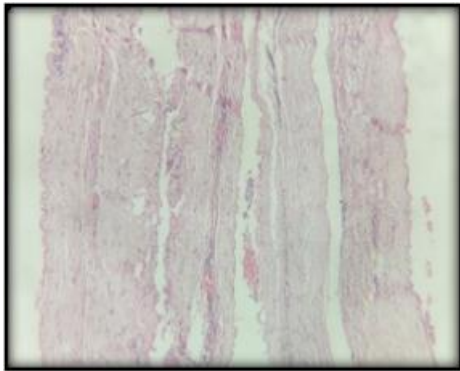


Figure 3:

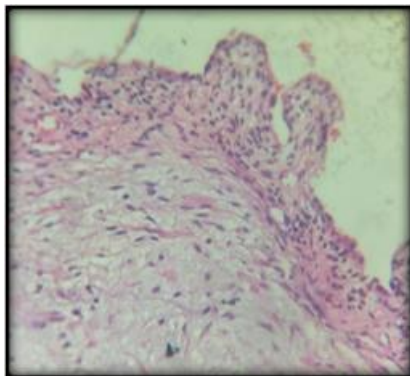


Figure 4:

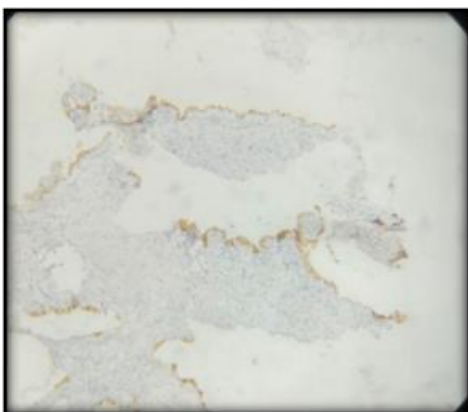


Figure 5: EMA: Positive in mesothelial cells

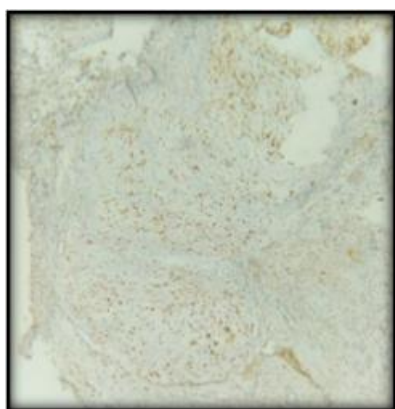


Figure 6: S100: Positive in solid area

3. Discussion

Pleuropericardial cysts are usually congenital but acquired causes such as inflammation, trauma, post cardiac surgery and

chronic haemodialysis have been reported. Congenital Pleuropericardial cysts are usually result from failure of fusion of one of the mesenchymal lacunae that form the pericardial sac, during embryogenesis after the third week of gestation.(1)

They are usually found in the third or the fourth decade of the life. Both sexes are affected equally. According to location 70% of the cases, these cysts are located in right cardiophrenic angle, in 22% cases in the left cardiophrenic angle and in 8% cases are located in the posterior or the anterior-superior part of the mediastinum(3). Most are asymptomatic but symptoms may occur because of compression and mass effect(4). Histopathologically a pericardial cyst has a simple wall of fibrous tissue lined by simple cuboidal mesothelial cells, without specialized epithelium or smooth muscle cells(4).

Differential Diagnosis

Differential diagnosis includes spectrum of mediastinal cysts, like bronchogenic cysts, enteric/esophageal duplication cysts, localized pleural/pericardial effusion, thymic cysts, cystic pericardial lymphangioma, and cystic teratoma. Routine microscopic examination with trichrome special stains may confirm the diagnosis in these cases(5).

Management

Management vary from conservative follow-up, percutaneous aspiration or surgical cyst excision by thoracotomy, sternotomy, or video-assisted thoracic surgery. Conservative treatment is usually applied in asymptomatic cases. Surgical excision is the “gold standard” therapeutic approach in symptomatic, complicated cases or atypical localizations, like cyst close to large vessels. The prognosis is excellent after resection, only one case of recurrence has been reported in literature(5).

4. Conclusion

Pleuropericardial cysts are rare and mostly they are clinically silent, but can occasionally cause life threatening complications. The majority of them are congenital due to developmental deficits and are most commonly found incidentally via routine radiography(1). Accurate diagnosis depends on various imaging techniques, and treatment varies from regular monitoring to surgery, depending on the symptoms and complications. With proper management, patients with pleuropericardial cysts usually have an excellent prognosis and quality of life. Ongoing research will shed light on the genetic and molecular causes, potentially leading to new diagnostic and therapeutic options(2).

References

- [1] Koumantzia C, Saridakis N, Eleftheriou A. Pleuropericardial Cyst: A Review of the Literature. *J Cardiol and Cardiovasc Sciences*. 2019 May 1;3(4):16–29.
- [2] Bhavsar DK. What Are Pleuropericardial Cysts? [Internet]. 2024 [cited 2024 Jun 19]. Available from: <https://www.icliniq.com/articles/respiratory-health/pleuropericardial-cysts>

- [3] Kar SK, Ganguly T. Current concepts of diagnosis and management of pericardial cysts. *Indian Heart J.* 2017;69(3):364–70.
- [4] Meredith A, Zazai IK, Kyriakopoulos C. Pericardial Cyst. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 [cited 2024 Jun 19]. Available from: <http://www.ncbi.nlm.nih.gov/books/NBK562287/>
- [5] Grigoraş A, Amălinei C, Căruntu ID, Grigoraş CC, Chiseliţă IR, Crişan-Dabija RA. Symptomatic pericardial cysts and dilemmas in their diagnosis. *Rom J Morphol Embryol.* 2023;64(4):517–25.