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(CASE REPORT)

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Pericardial mesothelial inclusion cysts versus chylothorax in a patient with high chest tube output postoperatively

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Abstract

Mesothelial inclusion cysts are rare and even more rare when encountered in and around the pericardium. This case report highlights the postoperative management of a patient with this rare disease, in combination with the development of a chylothorax after mitral valve surgery and the undefined relationship between the two disease processes in the setting of cardiofaciocutaneous syndrome.

Keywords: Mesothelial Inclusion Cyst; Chylothorax; Cardiofaciocutaneous Syndrome; Congenital Cardiac Surgery

1. Introduction

Mesothelial inclusion cysts are a type of benign tumor, lined by mesothelial cells, that are typically found in the peritoneal cavity and line the peritoneum[1]. They are rarely encountered in and around the pericardium. We present a case of multiple pericardial mesothelial inclusion cysts incidentally found intraoperatively in a patient undergoing mitral valve surgery. The postoperative course was prolonged due to high chest tube output and bilateral pleural effusions possibly due to chylothorax versus mesothelial inclusion cysts in a patient with known cardiofaciocutaneous syndrome. There are very few case reports in the literature highlighting multiple pericardial mesothelial inclusions cysts, and even less detailing the postoperative management of this rare finding after mitral valve surgery.

2. Case Report

A 9-year-old female with cardiofaciocutaneous syndrome with KRAS mutation and history of mitral valve regurgitation who previously underwent mitral valve repair surgery presented with persistent severe mitral regurgitation and severe left atrial dilation. She was scheduled to undergo redo mitral valve repair surgery. Upon opening the sternum, numerous cysts were encountered throughout the pericardium containing chylous appearing fluid. The cysts were excised and sent to pathology, which showed benign mesothelial lined tissue with reactive changes consistent with mesothelial inclusion cysts as can be seen in Figure 1.

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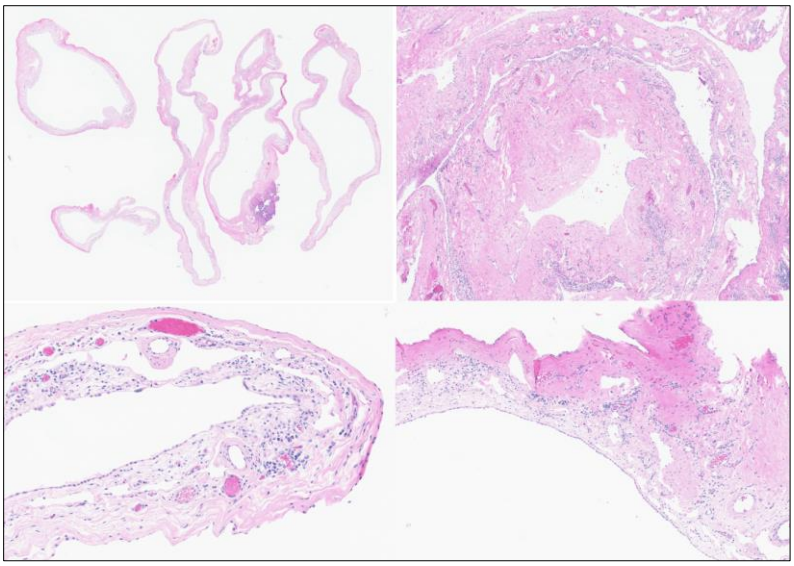

Figure 1 Histology showing benign mesothelial lined tissue with reactive changes

The postoperative course was notable for persistent high volume chest tube output that was milky in nature with elevated triglycerides. This was initially treated with a chylothorax protocol, which included a low-fat diet and octreotide with mild improvement in the chylous drainage. However, there was subsequent reaccumulation of the pleural effusions requiring diuretics on postoperative day 17 as seen in Figure 2. After aggressive titration of diuretics and maintenance of a low-fat diet, there was gradual improvement in the pleural effusions and resolution of the chylous drainage from the chest tubes which allowed for chest tube removal and discharge from the hospital on post operative day 41.

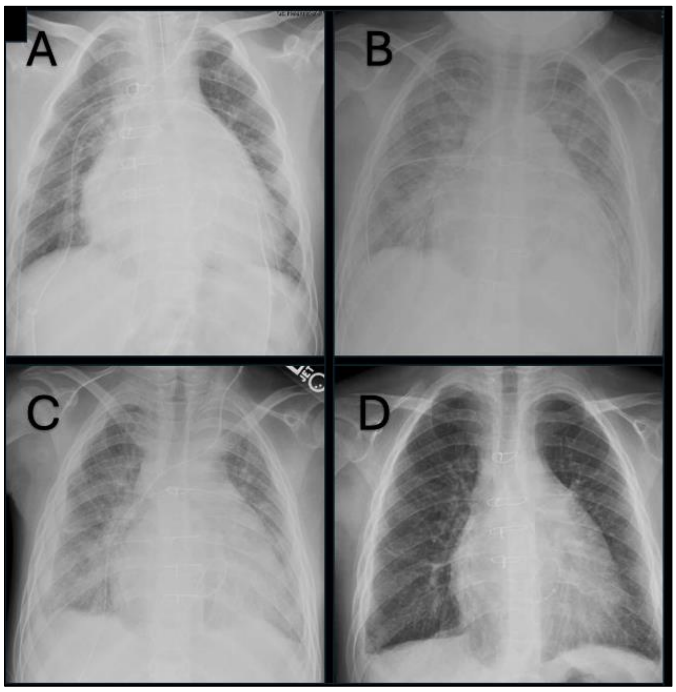

Figure 2 a) Postoperative day (POD) 0 CXR with mild worsening of scattered lung atelectasis and edema; **b)** POD 7 CXR with trace pleural effusions; **c)** POD 17 CXR showing increased right pleural effusion and trace left pleural effusion; **d)** POD 48 CXR (after discharge from the hospital) showing trace partially loculated bilateral pleural effusions

3. Discussion

Postoperative chylothorax may occur after congenital heart surgery with an incidence ranging from 2 to 5%[2]. The etiology may be secondary to traumatic injury to the thoracic duct intraoperatively or non-traumatic causes, as can be seen in several congenital syndromes[2]. Cardiofaciocutaneous (CFC) syndrome is a rare condition that is characterized by dysmorphic facial features, cardiac abnormalities, neurodevelopment delay, and lymphatic abnormalities such as lymphedema and, in isolated case reports, chylopericardium[3-5]. The case report by Akahoshi et al describes the incidental finding of chylopericardium in a 12-year-old female with a diagnosis of cardiofaciocutaneous syndrome with KRAS gene mutation[3]. This patient also had a prolonged hospital stay of almost 30 days and the chylopericardium only resolved after lymphangiography[3].

The RAS/mitogen-activated protein kinase (MAPK) pathway is a signaling cascade that is associated with cell proliferation, maturation, differentiation, as well as apoptosis[3,5]. Dysregulations in this pathway with genetic mutations in KRAS are known to cause various syndromes, including CFC syndrome[3,5]. However, the clinical manifestations of lymphatic dysplasia that occurs in patients with CFC and KRAS mutations has not been well documented and the pathophysiology between KRAS mutations and the observed lymphatic anomalies, such as chylothorax or chylopericardium, is not well understood[3,5]. Unfortunately, we do not have data to support or refute the role that this patient's KRAS mutation played in the development of her mesothelial inclusion cysts.

4. Conclusion

Mesothelial inclusion cysts are rare and even more rare when encountered in and around the pericardium. This case report highlights the postoperative management of a patient with this rare disease, in combination with the development of a chylothorax after mitral valve surgery and the undefined relationship between the two disease processes in the setting of cardiofaciocutaneous syndrome.

Compliance with ethical standards

Disclosure of conflict of interest

There are no conflicts of interest to disclose.

Statement of ethical approval

Ethical approval was obtained.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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