



Case Report

Chest wall cyst, a long term complication of pericardiectomy: A rare case report



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1. Case report

A 74-year-old male suffered from severe long standing rheumatoid arthritis (RA) and had been on long term Methotrexate and steroids. His condition complicated by recurrent pericardial effusion for which series of pericardial windows were done followed by partial pericardiectomy for constrictive pericarditis. Eighteen months later on, he presented with slowly progressive painless swelling at the lower end of the median sternotomy scar (Fig. 1). His concern was to exclude malignancy and not the disfigurement. Chest X-ray showed bilateral pleural thickening, and on lateral view, the mass was slightly lucent and resting on the lower end of sternum with surgical wires. Linear calcification was noted along cardiac apex and diaphragmatic copulae (Fig. 2). Ultrasound in the right parasternal region revealed a 6 cm avascular tense multi-septate complex solid cystic subcutaneous mass mostly containing proteinaceous material (Fig. 3). Contrast enhanced CT scan of the chest demonstrated a non-enhancing pear-shaped encapsulated cystic lesion measuring 5 × 8 cm in the subcutaneous tissue of the anterior chest wall and connected with the anterior mediastinum via small fistulous tract traversing a split in the sternum. Features of RA were evident in the form of: bilateral pleural thickening and calcifications, as well as bilateral pulmonary changes manifested as focal ground glass opacification, basal sub-pleural reticulation, and early cylindrical bronchiectatic changes; additionally, residual thin egg-shell pericardial calcifications along the inferior diaphragmatic surface of the pericardium was elicited as well as calcific spots at atrio-ventricular groove (Fig. 4). To the best of our knowledge, similar condition, post peri-cardiectomy chest wall cyst, has not been previously reported in the literature. In order to avoid any infectious complications of intervening inside a sterile cavity connected to the mediastinum/pericardial membrane, conservative management was undertaken and this was as well the favorable option for the patient; but he was instructed to seek medical advice once it render hot and painful, or when the

disfigurement starts to annoy him. Patient gave his signed consent and permission to the team of the Essex Cardiothoracic Centre to publish in any scientific journal the imaging material from his medical records and images obtained.

2. Discussion

The pericardium envelops the heart and the roots of the great vessels as a protective capsule. It is a cone shaped sac made up of two continuous and intimately connected layers, typically no more than 2 mm thick: (1) the inner thin visceral layer of the serous pericardium - the epicardium- is a transparent monolayer of mesothelial cells and forms part of the external surface of the heart; (2) the outer thicker parietal layer of the serous pericardium is a dense connective tissue and is adherent to the outermost fibrous pericardium. The in-between potential space contains normally no more than 50 mL of serous fluid. The pericardium serves two major functions: it maintains the heart position within the mediastinum and prevents cardiac distention by sudden volume overload [1]. Pericarditis, the most common pericardial disorder, has many etiologies including infectious (viral, bacterial, fungal), metabolic (uremic, drug induced), autoimmune (arthritis, thyroid), postradiation, neoplastic, traumatic, postinfarction (Dressler's syndrome, 10–15%), post-pericardiectomy (5–30%), and idiopathic. Nevertheless, a wide range of the fore-mentioned conditions promote pericardial scar formation, the pathologic process underlying constrictive pericarditis (CP) resulting in peri-cardial non-compliance and impeding the normal ventricular distensibility and diastolic filling [2]. For example, Rheumatoid arthritis, which is a chronic inflammatory disease of the small joints manifesting as pain, swelling and stiffness, and eventually joint destruction, can also be associated with extra-articular cardiovascular manifestations, the most common of which is the pericarditis. This is believed to be provoked by immune complexes precipitation in the pericardium and its diagnosis is often complicated

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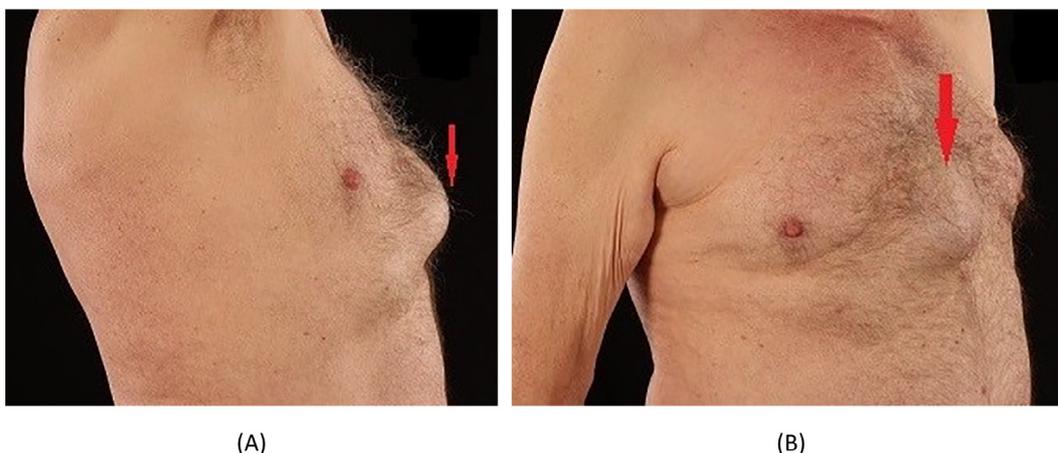


Fig. 1. (A and B) The patient complained of slowly progressive painless swelling at the lower end of the median sternotomy scar. No local signs of inflammation was evident (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

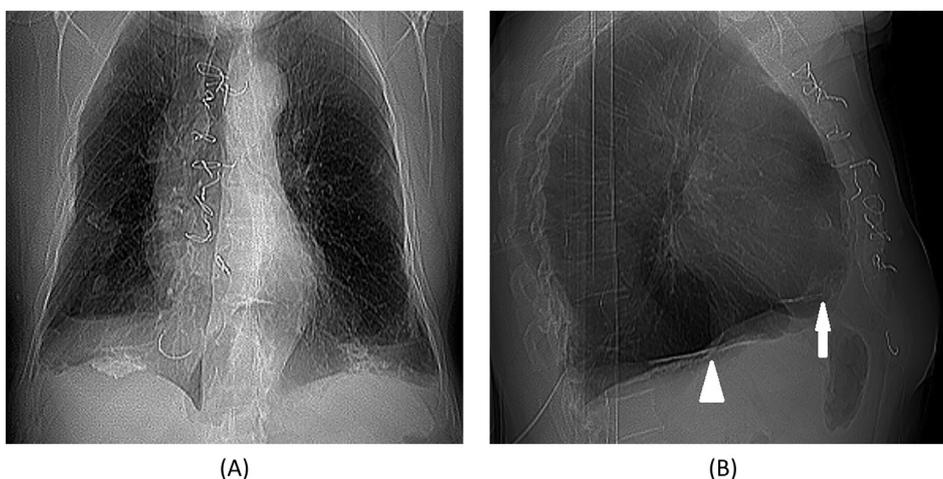


Fig. 2. Chest X-ray. Bilateral pleural thickening was noted in PA view (A), while pericardial and diaphragmatic pleural calcification are noted in lateral view (B).

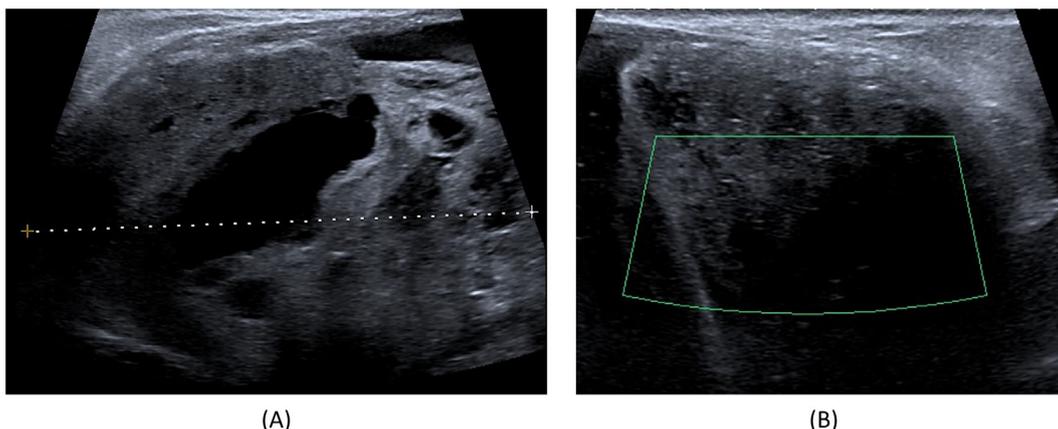


Fig. 3. (A and B) Ultrasonography excluded vascular herniation and revealed the complex nature of this multi-septated cyst.

by the many clinical variants and possible inter-current diseases such as drug-induced and viral pericarditis. Approximately half of patients with RA have pericardial effusions, and almost half of all patients with RA have significant pericardial adhesions at autopsy [2,3].

Clinically, the symptoms of pericarditis include chest pain (dull aching or tightness), weakness, malaise, fever (occasionally with rigors), cough or odynophagia; and when constriction develops, signs of right-sided congestion (jugular venous distention, edema, and ascites)

predominate, associated with high pulmonary capillary pressure and low cardiac output response to exercise resulting in dyspnea and effort intolerance [4].

Radiologically in CP, the chest x-rays (CXR) may show a characteristic arc shaped pericardial calcification. On the other hand, chest computed tomography (CT) is more superior in calcification detection plus it allows pericardial thickness measurement; moreover, magnetic resonance imaging (MRI) offers excellent accuracy (93%) in depiction

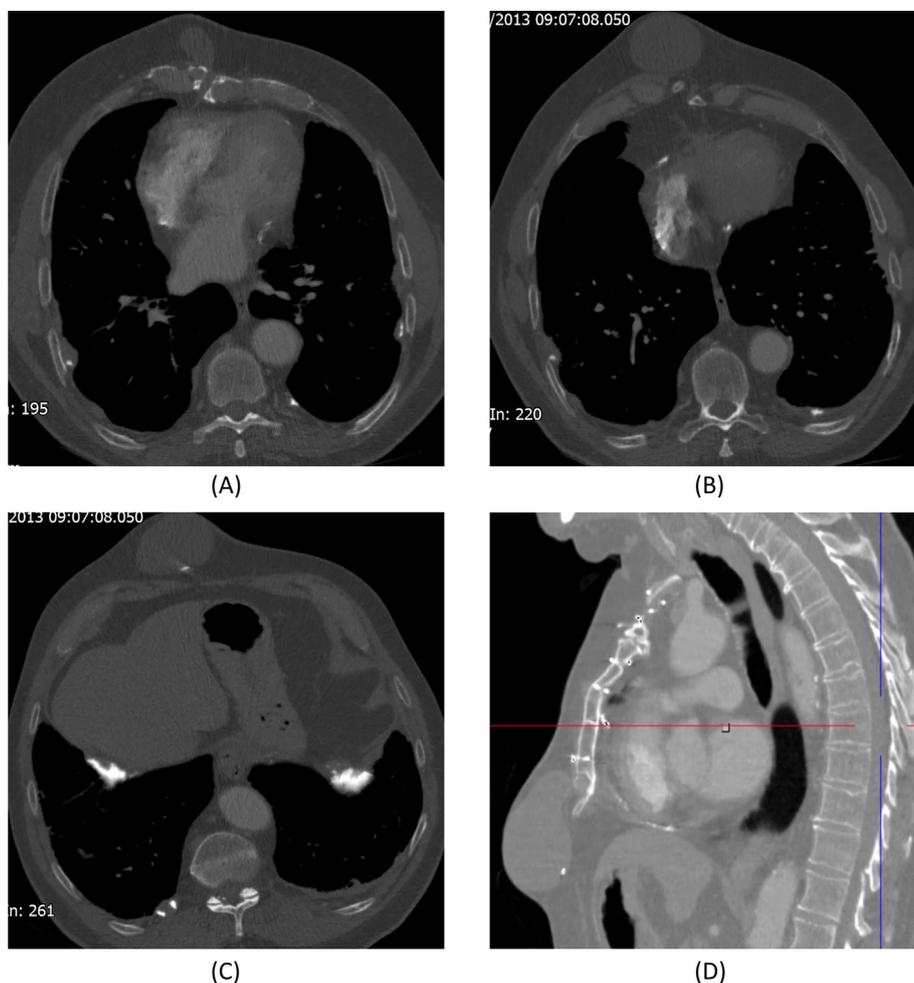


Fig. 4. CT scan with contrast injection showed: the sternal dehiscence through which the neck of the lesion was connected to the mediastinum as well as bilateral pleural thickening and calcification (A), the cystic nature of the lesion and calcified spots at atrio-ventricular groove (B), dense diaphragmatic copulae calcifications (C), and curvi-linear pericardial calcification.

of pericardial thickening > 4 mm, it may reveal coexisting active pericarditis which would alter the management plans, and may establish restrictive cardio-mayopathy when CP diagnosis is indefinite. Additionally, both CT and MRI allow the exclusion of non-cardiac causes of dyspnea and their dynamic protocols demonstrate many of the physiologic features seen echocardiography, even more reliably. However, it is always crucial to remember that around 18% of surgically proven CP cases have normal pericardial thickness [5].

Management of patients with pericarditis secondary to RA is variable according to its stage: pericardial drainage is often employed early for symptomatic effusions because of slow and doubtful response to medical treatment; on the other hand, pericardial window is created in cases with recurrent effusion through excision of a portion of the pericardium allowing the effusion to continuously drain into the peritoneum or chest. It is done either via subxiphoid incision, thoroscopically, or via a thoracotomy; and finally, pericardiectomy is considered for symptomatic relief of patients with CP in longstanding RA, in which stripping of the pericardium is done via median sternotomy [6,7]. However, all these surgical procedures carry potential risks and complications. In the literature, pericardial windows occasionally lead to a condition known as “pericardial hernia”, a type of diaphragmatic hernia where abdominal organs can herniate through the diaphragm and the pericardium into the pericardial space [8]. The reported post-pericardiectomy complications included pleural effusion, pulmonary infection, long-term intubation, low output syndrome (either due to associated cardiomyopathy or due to cardiac dilatation after

peri-cardiectomy), bleeding, acute renal failure, hepatic failure, and wound infection [9,10]. In contrast to these reported complications, a herniating cyst through the thoracic wall is a surprising complication post-pericardiectomy. The visible symptoms – an anterior chest bulge that may resemble malignancy in the eyes of the patient – can often be perplexing. It may be associated with a degree of malunion of the lower part of the sternum, which allowed the fistula to develop. Nevertheless, its development in a patient with severe rheumatoid arthritis (RA) raises the question of a possible link to inflammation. Pericardial defects have been known to occur in patients with severe inflammation; for example, in another acutely inflamed patient with severe deforming RA, an aorto-atrial fistula had spontaneously developed [11]. Interestingly, post-pericardiectomy syndrome (PPS) a common complication that occurs after cardiac surgery has been found to be preceded by a rise in pro-inflammatory and a decrease in anti-inflammatory serologic markers [12]. A recent double-blinded, randomized clinical trial showed that perioperative use of colchicine (a potent anti-inflammatory) reduced the incidence of PPS [13]. In our patient, it may have been the additive inflammatory effects of surgery and also of severe rheumatoid arthritis itself that caused a cyst followed by herniation through the thoracic wall.

Many disorders are presented as chest wall lump but many of them have characteristic radiological findings that help in differentiating one entity from another; hence it is crucial to be oriented with the various manifestations of these disorders for accurate diagnosis and optimal management [14].

3. Conclusion

Chest wall cyst may develop as a long-term complication of pericardiectomy and it should be suspected by physicians. Radiological imaging is fundamental to exclude any vascular structure herniation as well as malignancies, to reveal its cystic nature and to delineate the fistulous tract

Conflict of interest

None declared.

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