Recurrent constrictive pericarditis associated with seropositive rheumatoid arthritis: A case report

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ABSTRACT

INTRODUCTION: Constrictive pericarditis is an uncommon disease characterized by impaired diastolic filling of the ventricles, encased in a fibrotic pericardium resulting from an inflammatory process. Rheumatoid arthritis is a rare cause of constrictive pericarditis, usually due to a concomitant acute or chronic serositis.

PRESENTATION OF CASE: This paper presents a unique case of recurrent constrictive pericarditis associated with seropositive rheumatoid arthritis, requiring pericardiectomy and complicated three years later by recurrent fibrosis, demanding a second pericardiectomy.

DISCUSSION: Defining this entity as recurrent constrictive “pericarditis” might be a mistake, given that a total pericardiectomy was performed in first instance.

No risk factors for recurrent fibrosis have been identified in the current medical literature, apart from partial pericardiectomy. We cannot demonstrate exclusively that rheumatoid arthritis is a risk factor for recurrent pericardial/epicardial fibrosis due to the rare nature of the disease.

CONCLUSION: Further large scale studies are necessary to identify the risk factors for recurrence.

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1. Introduction

Constrictive pericarditis is characterized by impaired diastolic filling of the ventricles, encased in a fibrotic pericardium. Rheumatoid arthritis is an uncommon cause of constrictive pericarditis, usually resulting from effusive serositis.

Pericardiectomy is the only recognized curative treatment for constrictive pericarditis; relieving the constriction and enabling ventricular diastolic filling [1].

We present the first case in literature of recurrent constrictive pericarditis associated with seropositive rheumatoid arthritis, requiring a repeat pericardiectomy 3 years following the first event.

2. Presentation of case

A 64 year old lady presented in January 2006 with symptomatic effusive pericarditis (functional class NYHA III), with a background of poorly controlled rheumatoid arthritis (RA), positive to rheumatoid factor. The pericardial effusion coincided with a RA flare-up, treated initially with Naproxen and Colchicine.

The patient returned ten months later with worsening dyspnoea and global pericardial effusion, coinciding with an interruption of the therapy with colchicine for three months due to gastrointestinal side effects. Her symptoms improved with diuretics and the introduction of Rituximab for RA. Nevertheless tachycardia and elevated jugular venous pressure persisted, leading to the suspicion of constrictive pericarditis. Computed Tomography (CT) showed flattening of the interventricular septum (IVS) and the right ventricle, relating to constrictive physiology. Cardiac catheterization demonstrated equalization of the end-diastolic pressures and a typical dip-and-plateau ventricular waveform with preserved ventricular function.

Three months later new symptoms of heart failure arose and a transthoracic echocardiography (TTE) demonstrated global pericardial effusion, reduced volume ventricles and atrial flow reversal in hepatic veins. Respiratory variation measured 30% on mitral/tricuspid valve Pulsed Wave Doppler. Magnetic Resonance (MRI) depicted elongated compressed ventricles and diastolic displacement of the IVS at rest, exaggerated during inspiration, suggesting interventricular coupling [Fig. 1A].

Decision was made to relieve symptoms by aspirating the pericardial effusion, but a pericardiocentesis was unsuccessful, therefore an apical surgical pericardial drainage was accomplished, discharging 400 ml of haemorrhagic fluid. The pericardial cavity was partially obliterated by adhesions and pericardial biopsy showed marked fibrosis and infiltrates of macrophages and lymphocytes; no granulomas or malignancy were identified.

The ventricular morphology remained rather tubular as demonstrated on MRI along with pericardial thickening and abnormal septal motion with ventricular interdependence.

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Eventually in March 2008 a total pericardectomy was performed by an experienced surgeon, by dissecting the pericardium towards the right ventricle and the right atrium, down to the diaphragm and forward throughout the ascending aorta. A cardiopulmonary bypass (CPB) was established, but the aorta was not clamped. The excision was continued from the right towards the left phrenic nerve level. Histological examination of the pericardium showed fibrous thickening and lymphocytic infiltrates [Fig. 2A, B].

Substantial improvement in terms of symptoms and haemodynamics followed, as illustrated on TTE by the absence of respiratory variations in mitral/tricuspid inflow velocities and a normal size and inspiratory collapse of the inferior vena cava.

The patient remained fairly stable for three years until new onset of heart failure symptoms. Cardiac MRI suggested persisting constrictive physiology with reduced volume and elongated tubular
ventricles, pericardial thickening and IVS flattening during deep inspiration.

Upon these new findings repeat pericardiectomy was performed in March 2011 by the same surgeon with the aid of CPB. A 6 mm thick fibrous layer constricting the heart was dissected off, up to the phrenic nerves [Fig. 2C, D].

The second pericardiectomy produced a steady recovery with normal ventricular relaxation pattern on MRI [Fig. 1B] and absence of symptoms at 3 years follow-up.

3. Discussion

The most common aetiology of constrictive pericarditis is idiopathic or viral (42–49%), followed by previous cardiac surgery (11–37%), mediastinal irradiation (9–31%), connective tissue disorders (3–7%), including RA, post-infective (tuberculosis or purulent pericarditis in 3–6%) and other causes [2]. Tuberculosis is the main noxa in endemic countries [1].

Signs and symptoms are dominantly resulting from right heart failure, i.e. peripheral oedema, abdominal discomfort, ascites, jaundice and – in late stages – symptoms of elevated pulmonary pressure, despite normal left ventricular systolic function.

Several diagnostic instruments are available, but TTE remains the first line approach. Welch et al. [3] suggested diagnostic criteria including respiration-related IVS shift and reversal of expiratory diastolic flow in the hepatic veins. Cardiac catheterization is valuable especially in the differential diagnosis with restrictive cardiomyopathy: peculiar findings are the dip-and-plateau pattern of the right ventricle waveform and equalization of left and right end-diastolic pressures. Cardiac MRI provides clear pictures of IVS excision with interventricular dependence, ventricular area changes and quantification of pericardial thickness, effusion and calcification.

Anti-inflammatory medications may provide temporary relief in 10–20%, however the only accepted curative treatment is pericardiectomy [2]. This can be performed via median sternotomy, associated with higher risk of wound infection, or left anterolateral thoracotomy, with higher rates of respiratory failure [4]. Cardiopulmonary bypass may or may not be established, depending on presence of bleeding or tight adhesions [1]. Comparative studies between partial and total pericardiectomy showed lower mortality and reoperation rates with the latter [1–5].

Rheumatoid arthritis is a rare cause of constrictive pericarditis, developing from effusive serositis in coincidence with RA flares. This can be demonstrated by temporal link in our patient, furthermore a symptomatic relapse occurred on interruption of anti-inflammatory therapy and an improvement was brought by the introduction of Rituximab. This recurrent effusive pericarditis then slowly evolved into constrictive pericarditis, as demonstrated by fibrotic and inflammatory changes in the pericardial biopsies.

There are no characteristic histological findings in constrictive pericarditis associated with RA; usually non-specific inflammatory reaction with infiltrate of mononuclear cells, associated with deposits of fibrin is found. Another peculiar aspect of this case is the recurrence: defining this entity as recurrent constrictive pericarditis might be misleading, given that a total pericardiectomy was performed in first instance; nonetheless the clinical picture fitted the constrictive pathology. Partial pericardiectomy is the only documented risk factor for recurrence [5], therefore total pericardiectomy represents the sole preventative measure.

4. Conclusion

In conclusion, this is the first case in literature of recurrent constrictive pericarditis associated with RA requiring redo surgery. No risk factors for recurrent constrictive have been identified, apart from partial pericardiectomy and we cannot demonstrate that RA is a risk factor for recurrence, given the rarity of the association with constrictive pericarditis. Further evidences are necessary to identify the risk factors for recurrence.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

This case has been reported in line with the SCARE criteria [6].

Conflict of interest statement

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Ethical approval

Non applicable.

Author contributions

Votano D: study concept, data collection, writing the papers. Tsang GM: editing. Ashton-Key M: histological specimens reporting. Goboles L: editing.

Guarantor

Daniela Votano, Laszlo Goboles.

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