CASE REPORT

Delayed recurrent pleuropericardial effusions following atrial septal defect closure with Amplatzer septal occluder device: a case of postpericardiotomy syndrome?

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SUMMARY

The authors present the case of a 41-year-old woman, who presented with moderate pericardial effusion and bilateral pleural effusion 11 months following a secundum atrial septal defect closure with an Amplatzer septal occluder (ASO) device. The acute presentation responded well to diuretics and a short course of corticosteroid therapy. The patient, however, continued to experience pleuritic chest pain and a recurrence of pericardial effusion a month later. The patient showed significant symptomatic improvement with a 2-month course of down-titrating prednisolone. Six months later, the echocardiogram showed complete remission of pleural and pericardial effusion. The possibility of cardiac erosion following ASO implantation has been excluded. The aetiology of the reoccurrence of steroid-responsive pleuropericarditis remains unknown.

BACKGROUND

The use of a transcatheter device for the closure of atrial septal defects (ASDs) has become the method of choice in patients with suitable anatomy. Similar to surgical closure, transcatheter closure has the advantages of abolishing interatrial shunting and therefore reduces right heart volume load and the risk of paradoxical embolism. It also offers the advantage of being minimally invasive, thus lowering the risk of associated morbidity with open-heart surgery. The major sequelae of percutaneous device closure include cardiac erosion, perforation, arrhythmias, thrombosis, sepsis and device migration. Most of the complications with the use of Amplatzer septal occluder (ASO) in ASD closure occurred early after implantation, such as arrhythmias, sepsis and thrombosis. Cardiac perforation, erosion and displacement of the device have been reported as medium to long-term sequelae postimplantation (months to years).¹ Postprocedure pericardial effusion has been reported in previous studies, but commonly as an early complication during hospitalisation.² To the best of our knowledge, delayed recurrent pericardial and pleural effusions in the absence of device erosion or fracture as complications of the procedure has not been reported in the past. The only comparable report to our case was the manifestation of cardiac tamponade 5 years after

the placement of ASO, a sequel following the late erosion of the device.³

CASE PRESENTATION

ASD closure

A 41-years-old woman from Ghana presented to our cardiology clinic with a 6-month history of shortness of breath on exertion and occasional palpitations with changes in posture. She was previously fit and well, and participated in competitive hockey during her teenage years. She has a medical history of intermittent migraine and menorrhagia, in which she was taking the combined oral contraceptive pill for.

On examination, a right ventricular heave with a soft systolic murmur was noted. Initial bedside observations recorded the following: body mass index: 25.2, blood pressure: 153/86 mm Hg, heart rate: 72 bpm, SpO₂: 98% on air, with no elevated jugular venous pressure, no peripheral oedema and no radiofemoral delay. Her ECG showed normal sinus rhythm with a slightly rightward axis.

Her echocardiogram demonstrated a large secundum atrial septal defect measuring 28×20 mm with a left to right shunt, right heart dilatation in a situs solitus heart with normal cardiac connections.

The patient was admitted for ASD closure with a 32mm Amplatzer device (25 October 2013). The procedure was successful and she recovered without any immediate complications. Postprocedure echocardiogram demonstrated good systolic function, a dilated right ventricle with tricuspid regurgitation and a pulmonary arterial pressure of 25+10 mm Hg. She was discharged with a 6-month prescription of aspirin. Subsequent follow-up demonstrated a complete recovery; the device was well seated with no residual leak, no pericardial effusion, good biventricular function and no evidence of right ventricular volume loading.

First episode of pleuropericarditis

October 2014

Eleven months after the ASD closure, the patient presented to the hospital with a 3-week history of orthopnoea and lower chest pain radiating to the neck and epigastric regions. The pain was exacerbated by eating and lying flat. Taking non-steroidal anti-inflammatory drugs offered partial pain relief.

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Figure 1 CT coronary angiogram showing sagittal view of a well-situated Amplatzer device between the atria.

On physical examination, she had bibasal lung crepitations and leg oedema up to the knees; no pericardial rub could be heard.

Differential diagnoses based on clinical findings included post-pericardiotomy syndrome (non-specific pericarditis following cardiac procedure), device erosion, respiratory and gastrointestinal tract infections. Her ECG showed normal sinus rhythm with no ST segment changes and no evidence of acute pericarditis. Routine bloods were taken, showing negative troponin T (<3 ng/L), negative D-dimer and a raised C reactive protein (CRP) of >300 mg/L. Factor 7 was also noted to be low.

Further investigations were undertaken following her transfer to the cardiology ward. CT thorax showed a moderate pericardial effusion measuring up to 19 mm (with a Hounsfield unit of 15), making a diagnosis of haemopericardium less likely (see figures 1 and 2). Bilateral pleural effusions and lower lobe atelectasis or a partial collapse were also seen (see figure 3). The pulmonary trunk and main pulmonary arteries were dilated, possibly related to previous shunt or pulmonary hypertension. Moreover, a CT cardiac coronary angiogram was performed, with no evidence of coronary artery disease—the Amplatzer device was noted with no evidence of erosion; its position appeared to be satisfactory (see figures 1–4).

Figure 2 CT coronary angiogram showing a four-chamber view of the Amplatzer device and fluid within the pericardial sac.

Figure 3 A CT thorax image showing bilateral pleural effusions, larger on the left than the right. Associated left and right lower lobe atelectasis or partial collapse is also demonstrated. The Amplatzer device can also be visualised in this image.

Based on the elevated CRP, daily temperature spike and bilateral crepitation on the auscultation of the chest, a course of co-amoxiclav was commenced to cover for pneumonia. Following the acute management with diuretics and bisoprolol, she was discharged on a course of oral steroids to complete.

Subsequent episodes of symptomatic pericarditis

November 2014

Four weeks after discharge, the patient had another episode of sudden chest pain and shortness of breath. She had been experiencing fevers and had a blood pressure of 158/78 mm Hg. She was reassessed in cardiac outpatient clinic and was found to have a small pericardial effusion near the right ventricle and thickened pericardium on transthoracic echocardiogram. ECG showed non-specific abnormalities of ST and T wave in inferior and precordial leads.

Blood samples were taken demonstrating CRP >100 mg/L and erythrocyte sedimentation rate (ESR) 67. Creatine kinase and troponin were negative. Rheumatological causes were also considered, but her autoimmune screen was negative. She was also negative for hepatitis B core antibody, hepatitis C and T-spot test. Positive serological markers included cytomegalovirus (CMV) immunoglobulin (Ig)G, Epstein-Barr virus (EBV) IgG and herpes simplex virus (HSV) IgG.

Figure 4 An image showing the Amplatzer device between the atria on transoesophageal echocardiogram. The left disc of the Amplatzer device seals the atrial septal defect completely (top cavity) with no evidence of erosion or displacement.

As the pericarditis was steroid-responsive, a 7-day course of prednisolone 30 mg to be down-titrated over 6 weeks was initiated.

A discussion with the rheumatology team raised undifferentiated connective tissue disease as a possible differential diagnosis. Thus, she was followed up by both rheumatology and adult congenital heart disease teams over the subsequent weeks with good response to steroid therapy. She finally stopped steroidal treatment 2 months after the previous episode.

February 2015

The patient remained well for 3 weeks on completion of her steroids. In early February 2015, she started complaining again of the same dull ache on breathing and lying on the left side (3 months after the second episode). She self-medicated with prednisolone 20 mg, which she reduced progressively to stop after a week.

The echocardiogram taken after the week in which the symptoms started showed normal biventricular size, good systolic function and a well-sited ASO with no evidence of pericardial effusion. The pericardium remained to be of bright appearance as was noticed in previous echocardiograms. There was no evidence of a pleural effusion. ECG was reported as normal.

DIFFERENTIAL DIAGNOSIS

A number of differential diagnoses have been considered for her pleuropericarditis. Common causes of pericarditis include those of viral and autoimmune conditions. As a result, a viral screen has been performed. The results showed positive titres of IgG CMV, EBV and HSV. IgG titres are markers of past exposure; therefore, in the absence of any acute Ig markers and viral symptoms, her case is unlikely to be due to a virus. Moreover, the recurrent nature of the case also makes a viral aetiology less likely.

For an in-depth investigation for her case, our patient has been referred to rheumatology to rule out autoimmune explanations. Other than her pleuropericarditis, she was systemically well with no signs of weight loss or vasculitic symptoms. Coupled with her negative markers including antineutrophil cytoplasmic antibody and antinuclear antibody, a rheumatology cause was thought to be highly unlikely.

With infectious and rheumatological causes excluded, a diagnosis of pericardiotomy syndrome was made. This was based on the hallmark features she presented with. According to the 2015 European Society of Cardiology (ESC) guidelines, a diagnosis of postcardiac injury syndrome, which includes postpericardiotomy syndrome, may be reached if the patient has had cardiac injury plus two of the following hallmark features: (1) fever without alternative causes, (2) pericarditic or pleuritic chest pain, (3) pericardial or pleural rubs, (4) evidence of pericardial effusion or (5) pleural effusion with elevated CRP.⁴

Therefore, the diagnosis of postpericardiotomy syndrome applies to this case in a number of ways. Her clinical presentation of a low-grade fever (criteria 1), pleuritic chest pain (criteria 2), combined with echocardiographic evidence of pleural effusion and pericarditis (criteria 3) with a raised CRP (criteria 5) satisfied four out of the five criteria proposed by ESC.

TREATMENT

She has been treated with corticosteroids during acute flares of her presentation. A down-titrating regime has been adopted in which she has responded well to.

OUTCOME AND FOLLOW-UP

She remained free from recurrences of pericarditis for 18 months. An episode of chest pain was described in March 2016, but was thought to be due to acid reflux and responded well to antacids and simple analgesia. She reported to be pain free during her cardiology follow-up in October 2017.

DISCUSSION

In summary, our patient presented with two distinct episodes of pleuropericarditis; both episodes were responsive to steroids and were associated with an acute phase response. Considering her history of a transcatheter ASD repair, a suspicion of postpericardiotomy syndrome was raised.

Postpericardiotomy syndrome is a febrile condition characterised by chest pain, pericardial and pleural effusions, conventionally following cardiac surgery or myocardial infarction. The syndrome typically presents in the early postoperative period, typically within days or months. In the recent years, case reports have reported postpericardiotomy syndrome as a complication as a result of endovascular procedures, in particular coronary angioplasty.^{5 6}

The steroid-responsive nature of her pericarditis suggests an inflammatory cause for her clinical picture. Unremarkable results from her autoimmune screen enabled us to exclude rheumatological conditions commonly causing pleurisy, such as rheumatoid arthritis or systemic lupus erythematosus. Previous literature has documented the frequent co-existence of various viral agents in the presentation of postpericardiotomy syndrome. Thus, a theory of an autoimmune response accelerated by a viral infection has been proposed as a pathophysiology for this condition.⁷ Viral infections commonly associated with acute pericarditis include organisms typically causing upper respiratory tract infections; likewise, infections by cytomegalovirus may also present in this manner. In our case, the patient's blood culture results were negative. IgG was positive for CMV, EBV and HSV—this points towards the possibility of a past infection and is unlikely to hold significance in this case. The recurrent nature of the disease also reduces the likelihood of a viral aetiology.

Although we used the ESC guidelines as a basis for our diagnosis, it is worth noting that ASO insertion has not been included in the guidelines as an example of 'cardiac injury'. Nevertheless, since coronary percutaneous intervention and pacemaker lead insertion have been included as examples of iatrogenic cardiac trauma, extrapolation of the principle may be appropriate in this case.

The delayed onset of pericarditis to almost up to a year since the procedure makes this case an atypical case of postpericardiotomy syndrome. To our best of knowledge, recurrent pleuropericarditis following ASO implantation has not been reported before and thus, makes this case highly unique.

Learning points

- The awareness among physicians for pericardiotomy syndrome following a transcatheter device closure of an atrial septum defect as a late complication should be raised.
- Pericarditis following cardiac injury can follow a relapsing course; thus, regular follow-up is essential.

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