Novel treatment (new drug/intervention; established drug/procedure in new situation)

Recurrent chylopericardium

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Summary

A 19-year-old male presented with recurrent episodes of pericardial effusion. On evaluation, he was diagnosed to have idiopathic chyloous effusion. As there was persistent pericardial collection, chemical pericardiodesis was done. Following pericardiodesis, there was no recurrence of pericardial effusion. Chemical pericardiodesis may be an effective treatment option for recurrent, idiopathic chylopericardium.

BACKGROUND

Chylopericardium is a rare condition characterised by accumulation of triglyceride rich fluid in the pericardial cavity. Though several etiological conditions have been described, it can be a primary idiopathic disorder.¹ Treatment of idiopathic recurrent chylopericardium is difficult and surgery has been recommended for refractory patients.² We present a case, which demonstrates that chemical pericardiodesis may be a useful alternative to surgical treatment in this scenario.

CASE PRESENTATION

A 19-year-old male presented with breathlessness, which had progressed from New York Heart Association class II to IV, over a period of 1 month. There was no history of chest pain, joint pain, fever or respiratory tract infection. He had loss of appetite but there was no significant weight loss. He was diagnosed to have pericardial effusion and had undergone pericardiocentesis twice within the past 2 years. He was advised to take antitubercular drugs empirically during the last episode of pericardial effusion. The nature of the pericardial fluid during those episodes was not known. He was not a known diabetic or hypertensive. There was no history of chest injury or thoracic surgery.

On admission, he was tachypnoeic. His heart rate and blood pressure were 124/min and 90/70 mm Hg, respectively. There was no pedal oedema or lymphadenopathy. The precordium was unremarkable and the lung fields were normal on auscultation.

INVESTIGATIONS

The complete haemogram, white blood cell count and erythrocyte sedimentation rates were normal. The blood sugar, renal function tests, liver function tests and thyroid profile were also within normal limits. The total cholesterol level was 184 mg/dl, high density lipoprotein, low density lipoprotein and triglycerides were 58, 107 and 90 mgs/dl, respectively. Serology for HIV, hepatitis B surface antigen, hepatitis C virus, Venereal Disease Research Laboratory and antinuclear antibodies were negative. His

electrocardiogram showed sinus tachycardia and chest x-ray was suggestive of pericardial effusion. An emergency echocardiogram revealed a large pericardial effusion with cardiac tamponade and immediate pericardiocentesis was done. The cardiac chambers and valves were structurally normal with adequate biventricular function. About 1200 ml of milky white pericardial fluid was aspirated and analysed. The composition of pericardial fluid is enumerated in table 1.

These findings were consistent with the diagnostic criteria for chylopericardium² that is milky white aspirate, triglyceride >500 mg/dl, cholesterol/triglyceride ratio <1.0, negative culture and preponderance of lymphocytes. A CT of chest (figure 1) showed normal thoracic structures. There was no evidence of lymphadenopathy or mass lesion and the heart was structurally normal with moderate pericardial effusion. The pericardial thickness was normal on CT imaging. Pericardial biopsy demonstrated non-specific inflammatory cells and there was no evidence of tuberculosis or malignancy (figure 2).

DIFFERENTIAL DIAGNOSIS

Tuberculous and malignant effusions are the most common differential diagnosis of recurrent pericardial effusion. In this patient, both tuberculosis and malignancy have been ruled out by appropriate investigations. Hence this is a case of idiopathic recurrent chylopericardium.

Table 1 Pericardial fluid analysis.

Pericardial fluid	
Total count	1900 cells/mm ³ (predominantly lymphocytes no malignant cells)
Protein	5 gm/dl
Sugar	145 mg/dl
Total cholesterol	90 mg/dl
Triglyceride	1186 mg/dl
Cholesterol/triglyceride ratio	0.07
Gram stain/AFB stain	Negative
Bacterial/Fungal culture	No growth
PCR for Tubercule bacilli	Negative

AFB, Acid fast bacteria

Figure 1 CT scan of thorax demonstrating moderate pericardial effusion. There is no hilar or mediastinal tumour or lymphadenopathy.

Figure 2 Eosin haematoxylin staining of pericardial biopsy specimen revealing non-specific inflammatory cells. The absence of caseating necrosis or malignant cells can be appreciated.

TREATMENT

As there was continuous pericardial drainage, pericardiostomy was done. There was drainage of 500–600 ml of fluid/day. Following initiation of low fat diet, enriched in medium chain triglycerides, the drain decreased to 50–60 ml/day. As patient was not willing for surgery, he was given the option of chemical pericardiodesis, to which he consented. Two grams of oxytetracycline injection mixed with 100 mgs of lignocaine was instilled in the pericardial cavity and the drain was clamped. The patient's position was continuously changed and the fluid was drained after 2 h. The drain was left insitu for 48 h. As there was no further collection of pericardial fluid, the drain was removed.

OUTCOME AND FOLLOW-UP

Further in hospital course was uneventful and he was discharged. He is asymptomatic at 1.5 years of follow-up with no evidence of constrictive pericarditis or recurrence of pericardial effusion.

DISCUSSION

Idiopathic chylopericardium is an uncommon condition, which is diagnosed by excluding specific etiological conditions like malignant tumours, blunt or penetrating trauma, postcardiac surgery, infection, radiation, gastric signet ring cell carcinoma, Gorham syndrome, congenital lymphatic anomalies and congenital mediastinal lymphangiectasia.² CT thorax is essential to rule out tumours and other intrathoracic conditions. Though lymphoscintigraphy is useful to identify possible fistulous connection between thoracic duct and pericardial space, it is not readily available.

The possible mechanism of accumulation of chylous fluid includes presence of damaged lymphatic channels that cause efflux of chyle from thoracic duct to pericardial space and elevated pressure in thoracic duct which may occur in lymphangiectasia. Idiopathic chylopericardium may remain asymptomatic or it may have a subacute presentation. Rarely, it manifests as cardiac tamponade as described in this case.

Treatment option for chylopericardium includes conservative therapy or surgery. The indications for conservative therapy are first episode of chylopericardium, patients at high surgical risk and when patient's life span is limited by comorbid conditions. This includes pericardiocentesis followed by pericardiostomy together with a low fat, medium triglyceride rich diet. This conservative approach has a failure rate of 57.1%.³

Surgery is invariably curative for idiopathic chylopericardium. It is recommended when the pericardial drainage

exceeds 1500 ml/day or when it is more than 500 ml/day over a period of 5 days or when there is recurrence.² Surgery involves ligation of thoracic duct and creation of pericardial window with or without partial pericardiectomy.⁴ The drawback of surgery is that it is invasive and adds to the procedural cost.

Chemical pericardiodesis has been used successfully to treat recurrent pericardial effusion due to malignancy.⁵ This treatment option has not been tried in patients with recurrent chylopericardium. It is a simple, relatively non-invasive procedure. The tetracycline induced pericardial sclerosis prevents accumulation of fluid. The risk of future constrictive pericarditis is negligible. Instillation of tetracycline can be repeated till the pericardial drain decreases to 25 ml/day. Tetracycline instillation can be given up to a maximum of 5 cycles. In our patient, a single instillation was sufficient to induce pericardial sclerosis. The patient had no evidence of constrictive pericarditis at 1.5 years of follow-up. Chemical pericardiodesis is a simple, safe and cost effective alternative therapy to surgery for idiopathic recurrent chylous effusion.

Learning points

- ▶ Chylous pericardial effusion, though uncommon, should be considered in the differential diagnosis of recurrent pericardial collection.
- ▶ Life threatening cardiac tamponade may be the initial manifestation of chylopericardium.
- ▶ Detailed evaluation is mandatory to exclude secondary causes of chylopericardium.
- ▶ Chemical pericardiodesis may be a useful alternative for idiopathic chylopericardium, in patients unwilling for surgery.

Competing interests None.

Patient consent Obtained.

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