A case of haemorrhagic pericardial tamponade in an adolescent

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Pericardial effusions are occasionally encountered, and can be of infections, autoimmune, malignant and idiopathic aetiology. Large pericardial effusions may result in cardiac tamponade, which is a medical emergency. We report a case of a massive haemorrhagic pericardial effusion complicated by tamponade in a nineteen year old chef apprentice. He underwent an emergency pericardiocentesis, and made a quick recovery with symptomatic management. Upon follow-up, there was no recurrence of his effusion, and after extensive analyses of the fluid, no clear aetiology could be determined. Idiopathic pericardial effusions often pose a management challenge due to the difficulty of predicting the natural course and risk of recurrence.

Introduction

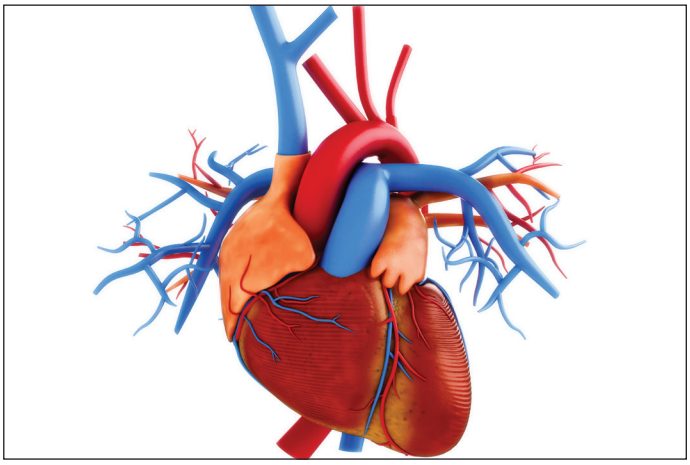
Pericardial effusion is the presence of excessive and sometimes abnormal composition of fluid in the pericardial space. Conditions that injure or insult the pericardium may lead to a pericardial effusion. In up to 60% of cases, it is associated with an identified or suspected underlying process and often linked with inflammation of the pericardium. [1] Nevertheless, in many cases, the underlying cause cannot be identified after extensive evaluation. Management of these idiopathic cases is more difficult due to their less predictable clinical course. To complicate the management, the patient in this case has haemorrhagic pericardial tamponade. Malignancy and tuberculosis are causes of haemorrhagic pericardial effusion that must be ruled out.

The case

A 19 year old male, working as a chef apprentice, presented to the emergency department with acute onset of pleuritic chest pain and a two week history of progressive shortness of breath. The pain was characteristically sharp, central and aggravated on inspiration and supine position. He was systemically unwell with chills and night sweats. There were no prodromal respiratory tract symptoms, palpitations, syncope, cough, sputum and wheeze. He was otherwise healthy. He denied engaging in any high risk behaviour, any sick contacts or travel to the tropics.

On physical examination, temperature was 37.7°C, respiratory rate 20 breaths/minute, heart rate 114 beats/minute and BP 136/78mmHg. Oxygen saturation was 94% on 3L of oxygen. Cardiovascular examination was remarkable for distended neck veins, pulsus paradoxus of up to 20 mmHg and muffled heart sounds on auscultation. He had normal vesicular breath sounds over all lung fields. There was no lymphadenopathy or palpable masses to suggest malignancies, and no localizing signs to suggest a focus of infection.

An emergency echocardiography demonstrated a large pericardial effusion with right atrial and ventricular diastolic collapse, suggestive of cardiac tamponade. His chest X-ray revealed an enlarged cardiac silhouette and a small right pleural effusion.



An urgent pericardiocentesis was performed and 600mL of haemorrhagic fluid was drained through a pig tail catheter, with instantaneous improvement of his symptoms. Fluid analysis was consistent with an exudative effusion as determined by Light's criteria. However, focused evaluation for infective aetiology including viral serologies, serology for atypical organisms and mycobacterium were negative. No malignant cells were identified in the fluid. Table 1 reflects the extensive evaluation that was performed.

After a 48-hour period, his drain was removed. He was discharged home with a 6-week course of indomethacin for the intermittent pleuritic pain that persisted for a further two weeks with pantoprazole for gastroprotection. Upon subsequent follow-up a week later, there was no recurrence of his effusion with full resolution of symptoms. The repeat echocardiography performed a month later was normal.

Discussion

Recognizing pericardial tamponade

The pericardial space can hold approximately 15-50 mL of fluid under normal circumstances. The pericardial fluid acts as a lubricant between the parietal and visceral layers of the pericardium. This fluid is believed to be an ultrafiltrate of the plasma produced by the visceral pericardium. When significant amount of pericardial fluid accumulates, it develops into pericardial effusions. Large effusions may contain greater than 2 litres of fluid. [2]

Pericardial effusions can be classified according to its time course, and the duration of its development influences clinical symptoms and presentation. This patient, who was previously healthy, developed acute symptoms of chest pain and dyspnoea due to rapid accumulation of 600mL fluid over 2 weeks. On the contrary, if the fluid is accumulated slowly over months, it allows the pericardium to stretch and adapt, and hence the patient can be asymptomatic. [3]

The morbidity and mortality of pericardial effusion is determined by its aetiology. The aetiology is typically established by the evaluation

Fluid Analysis	Exudative fluid • Effusion protein/serum protein - 56/57 = 0.98 (>0.5) • Effusion LDH/ serum LDH - 1835/238 = 7.71 (>0.6) • Effusion LDH - 1835U/L → >2/3 of upper limit of normal serum LDH Cytology: Predominantly neutrophils. No malignant cells. No growth. Immunohistochemistry assessment: Negative
Blood Culture	No bacterial growth.
Autoimmune screen	ANA, ANCA, C3, C4, Rheumatoid factor, ENA antibodies, anti-DNA, CCP – Negative
Viral serology & nasal swab PCR	RSV, rhinovirus, metapneumovirus, adenovirus, parainfluenza, influenza A and B, CMV, enterovirus, coxsackie virus – Negative
Bacterial serology, nasal swab and urine PCR	*Bordetella pertussis, Legionella pneumophila, Mycoplasma pnuemoniae, Chlamydia trachomatis, Neisseria gonorrhoeae, Treponema pallidum, Streptococcus pneumoniae:* all negative
Mycobacterium culture	For tuberculosis – Negative

Table 1. *Various investigations performed to evaluate the cause of effusion.*

of fluid analysis in relation to the clinical context in which it occurs. Most patients tolerate acute idiopathic effusions well, and have an uncomplicated recovery. In patients with tuberculous pericardial effusions, the mortality is 80 to 90% if left untreated. The mortality is reduced to 8% to 17% with anti-tuberculosis medication. [4] On the other hand, symptomatic effusion is one of the contributing cause of death in 86% of cancer patients with malignant effusions. [5] Conversely, for patients with large, chronic effusions lasting longer than 6 months, up to 50% of them can be asymptomatic. [6]

Cardiac tamponade is one of the most fatal complications of pericardial effusion. Clinically, it can be recognised from Beck's triad of muffled heart sounds, increased jugular venous pressure, and pulsus paradoxus. Our patient had a significant paradoxus of 20mmHg (normal <10mmHg) and elevated jugular venous pressure that made us suspect tamponade on clinical grounds.

In tamponade, increased intrapericardial pressure compromises ventricular filling and reduces cardiac output. This tamponade physiology exaggerates the typical respiratory variation in left and right ventricular filling, which explains the pulsus paradoxus. The time frame over which effusions develop determines the risk of developing a tamponade. An acute accumulation as low as 150ml can result in tamponade. [3]

Echocardiography is essential in the work-up of a patient with pericardial effusion. It demonstrates the size and presence of an effusion, which is visualised as echo-free space (Figure 1). However, the size of the effusion cannot accurately predict the possibility of cardiac tamponade. Cardiac tamponade is a clinical diagnosis. The general rule is pericardial effusions causing tamponade are usually large and can be seen both anteriorly and posteriorly. Other suggestive echocardiographic features of tamponade are right atrial collapse, and right ventricular collapse (Supplementary Figure 2). The sensitivity of identifying right atrial collapse for the diagnosis of tamponade is 50 to 100%, whereas the specificity is 33 to 100%. The sensitivity and specificity in finding right ventricular collapse ranges from 48% to 100% and 72% to 100% respectively. These findings by themselves are unreliable signs of tamponade clinically. They only have diagnostic value if the pre-test likelihood of tamponade is high for the patient in question. [7] It is hard to differentiate haemorrhagic and serous effusion on echocardiograph. However, fibrinous strands on echocardiograph suggest that an inflammatory process is present in the pericardial space, which was seen in this patient's echocardiogram. [8] Hence, the initial suspicion for this healthy young man was cardiac tamponade caused by viral pericarditis.

Significance of haemorrhagic pericardial effusion

Based on the suspicion of viral pericarditis, it was foreseen that the

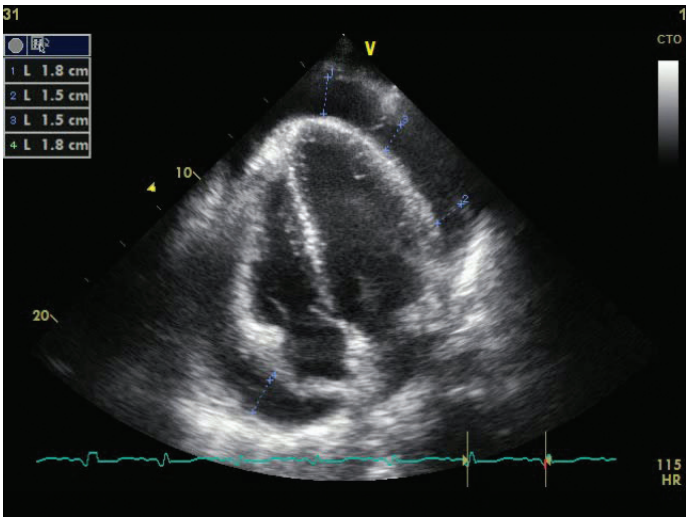


Figure 1. *Pericardial effusion in this patient as echo-free space.*

Video link: https://vimeo.com/133111472

Figure 2. *Video available online*

effusion would be serous. However, it turned out to be haemorrhagic pericardial effusion, which altered the diagnostic and management pathway.

The aetiology of pericardial disease is best categorised based on inflammatory, neoplastic, vascular, congenital and idiopathic causes. It has been noted that a definite cause for pericardial effusion has only been found in 60% of the patients. [1]

There have been major analysis studies addressing the issue of diagnosing and managing large pericardial effusions of unknown origin, [1,9,10] but only one study has discussed the aetiology of large haemorrhagic pericardial effusion. [11] Atar et al's study [11] evaluated 96 cases of haemorrhagic pericardial effusion and highlighted the common causes: iatrogenic (31%), malignancy (26%), postpericardiotomy syndrome (13%), idiopathic (10%). Traditionally, malignancy and tuberculosis have always been considered as potential causes. [2] However, as reflected in Atar et al's study, the incidence of tuberculosis has decreased and there is a rise in cardiovascular procedures over the past decade, resulting in a switch to iatrogenic disease as a major cause. Although it has been known that viral pericarditis can cause haemorrhagic effusions in rare cases, the frequency is unknown.

In our patient, extensive testing was performed to rule out common causes of pericardial effusion. However, no specific cause could be identified and the diagnosis of idiopathic pericardial effusion was made. In patients with idiopathic pericardial effusion, the aetiology is often presumed to be viral or of autoimmune cause. The proliferation of an infective agent and release of toxins can injure the pericardial tissue, causing haemorrhagic inflammation. Additionally, the pericardial involvement in systemic autoimmune conditions is thought to be due to the dysfunction of the innate immune system. [6] His low grade fever, exudative pericardial fluid, neutrophilia, and absent growth in the fluid culture supported the postulation of a viral cause.

Management of idiopathic pericardial effusion

The indications for pericardiocentesis are pericardial tamponade and for effusions more than 20mm, measured in diastole on echocardiograph. [12] When pericardial effusion is associated with pericarditis, management should follow that of pericarditis. The mainstay of therapy for patients with idiopathic pericarditis is nonsteroidal anti-inflammatory agents (NSAIDs), which is aimed at symptom relief. It has been shown that NSAIDs are effective in relieving chest pain in 85 to 90% of patients. [13] While colchicine is the definitive treatment for relapsing pericarditis, a limited number of small trials have also suggested that colchicine alone or in combination with NSAIDs can prevent recurrences when used in the first episode of acute pericarditis. Glucocorticoids should only be used in patients with contraindications or are refractory to NSAIDs and colchicine. [14]

The outcome of patients with large haemorrhagic pericardial effusions is dependent on the underlying disease. The mean survival for patients with malignant pericardial effusion was 8 ± 6 months post pericardiocentesis. In contrast, patients with idiopathic pericardial effusion have a favourable survival outcome similar to the general population. [11] Although no patients had recurrent effusion subsequently in Atar et al's study, it is known that with acute idiopathic pericarditis, there is a 10-30% chance of developing recurrent disease, and often with an effusion. A single recurrent attack may happen within the first few weeks after the initial attack, or as repeated episodes for months. [15,16] The pathogenesis of recurrent pericarditis is unclear, but has been speculated to be due to an underlying autoimmune process. [16] With recurrent episodes, the repeated inflammation can lead to chronic fibrotic scarring and thickening of the pericardium, resulting in constrictive pericarditis. [6]

There is no specific feature that reliably predicts the recurrence of idiopathic effusions. However, it has been shown that patients who responded well to NSAIDs have a lesser chance of recurrence, [17] while initial treatment with corticosteroids favours occurrence of recurrences due to deleterious effect on viral replication. [18] This patient had a good response to NSAID within a week with improvement in his symptoms, which subsequently fully resolved. This further supports the diagnosis of idiopathic pericarditis and is also a good indicator that he is not at an increased risk of recurrence. However, it would be beneficial for this patient to be reviewed in the future with repeat echocardiography if clinically warranted.

Conclusion

Cardiac tamponade is a life-threatening medical emergency that requires prompt diagnosis and emergent treatment. It is essential for one to be able to recognize Beck's triad. Haemorrhagic pericardial effusion is a red flag that warrants a meticulous search for uncommon but sinister aetiologies, especially malignancy and tuberculosis, as the mortality rate is high if left untreated. When extensive investigations have been conducted and the diagnosis of idiopathic pericarditis is made, NSAIDs are the mainstay of therapy. Colchicine can be considered to prevent recurrence, while glucocorticoids should only be used as a last resort.

Consent declaration

Informed consent was obtained from the patient for publication of this case report and accompanying figures.

Acknowledgements

None.

Conflict of interest

None declared.

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