

**CASE REPORT****A VEXING RECURRENT PERICARDIAL EFFUSION WITHOUT UNDERLYING PATHOLOGY – SUBSEQUENT DIAGNOSIS OF LYMPHOMA PROVES A RARE FIRST PRESENTATION****Haseeb Ur Rahman, Saad Hasan, Stephen Hutchison**

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An elderly man had recurrent admissions with large symptomatic pericardial effusions. Initial computed tomography (CT) of thorax, abdomen and pelvis and pericardial fluid analysis did not reveal underlying cause. On subsequent presentation, pericardial window was formed but repeat pericardial fluid analysis and biopsy failed to give a diagnosis again. He then presented approximately after four months with worsening symptoms of dyspnoea and weight loss. General physical examination at that point noted inguinal lymphadenopathy. Repeat imaging with CT and magnetic resonance imaging (MRI) showed features of metastatic malignancy. Tissue diagnosis from inguinal lymph nodes proved to be diffuse large B-cell lymphoma (DLBCL).

**Keywords:** Pericardial effusion; Inguinal lymphadenopathy; Diffuse large B-cell lymphoma

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**CASE REPORT**

A 71-year-old patient was referred to cardiology clinic by general practitioner with few weeks' history of insidious onset dyspnoea on exertion without chest pain. He had a background history of asbestos related pleural plaque disease. He was not on any regular medications. His physical examination was within normal limits. Chest X-ray showed large globular heart. Echocardiography (ECHO) confirmed large pericardial effusion with right ventricular, right atrial and left atrial diastolic collapse. Inferior vena cava (IVC) diameter on expiration was 2.4 centimetre (cm) while inspiratory IVC diameter was 1.9 cm.

He was admitted to the hospital to investigate the cause of pericardial effusion. Full blood count, C reactive protein and serum biochemistry was unremarkable. Pericardiocentesis was performed and 1500 millilitres (ml) of straw-coloured pericardial fluid was drained which was sent to the lab to be tested for cell count, biochemistry and pathology/cytology. The results showed raised protein of 58 gram/litre (g/L), cell count showed 100% lymphocytes and gram stain and Ziehl-Neelson stain for bacteria and mycobacterium respectively were also negative. Serum protein was noted to be 78 g/L. Cytology showed no malignant cells. CT of thorax, abdomen and pelvis was performed and was unremarkable.

A follow up outpatient ECHO at 8 weeks showed recurrence of large volume pericardial effusion with echocardiographic cardiac tamponade features. Despite it, he was well. A repeat pericardiocentesis was performed. Samples tested in the lab were no different than previously. He was then referred for pericardial window to cardiac

surgeons in the tertiary centre. Pericardial biopsies were performed there but the results did not explain any cause of pericardial effusions.

Three weeks later, he returned to Accident and Emergency department with worsening shortness of breath, weight loss and dizziness. General physical examination on this admission was remarkable for new inguinal lymphadenopathy. ECHO was repeated that showed recurrence of moderate volume pericardial effusion. A subsequent CT scan was planned on suspicion of the pericardial effusion being loculated which showed very aggressive tumour lesion affecting both pleurae associated with bony tumour infiltration at the thoracic spine level and bilateral mediastinal tumour infiltration extending and invading the superior pericardial recesses surrounding the aorta. There was encasement of the aortic arch, and of the superior vena cava (SVC). Bilateral nodal mediastinal and hilar disease was also seen. MRI spine findings raised suspicion of bony metastatic disease.

Biopsies were taken from inguinal lymph nodes that revealed the diagnosis of DLBCL. Haematology input was taken and he was referred for aggressive chemotherapy.

It took about three months and twenty days from first presentation of pericardial effusion with no evidence of underlying pathology to presenting again with B symptoms with clinical and radiological features of lymphoma confirmed with a tissue diagnosis.

**DISCUSSION**

We describe a rare case of puzzling pericardial effusion which on early thorough investigations was

considered to be idiopathic. However, eventually patient developed B-symptoms with clinical and radiological lymphadenopathy. Histological diagnosis was confirmed as DLBCL. Unexpectedly, it took 4 months from initial presentation with pericardial effusion to actually developing the clinical and radiological features of the underlying lymphoma.

Cardiac involvement is usually observed quite late in lymphoma. It takes a mean of 20 months after diagnosis and often found at autopsy.<sup>1</sup> Interestingly, DLBCL is the most commonly reported histologic subtype of cardiac non-Hodgkin's lymphoma (NHL)<sup>2</sup> that was also the diagnosis in this patient. Overall, it is still uncommon to have symptomatic cardiac involvement in patients diagnosed with NHL. Hence, there is very limited evidence which have measured patient outcomes.<sup>2</sup>

The most common causes of pericardial effusion with hemodynamic compromise are malignancy (lung, breast and melanoma) followed by infections, iatrogenic injury, trauma, metabolic causes, uraemia, and connective tissue diseases.<sup>3</sup>

Symptomatic patients who present with shortness of breath, chest pain, palpitations with clinical signs of pericardial disease like muffled heart sounds, raised venous pressure, pericardial rub or hemodynamic compromise are investigated by an ECHO. Size of pericardial effusion is determined by echo free space between myocardium and pericardium in diastole in different views. Effusion can be categorized into large >20 millimetre (mm), moderate 10–20 mm and small <10 mm. However, only the size of pericardial effusion does not determine if it would have hemodynamic effects. Even a small pericardial effusion that builds up quickly can cause cardiac tamponade.<sup>4</sup>

Cardiac tamponade is characterised by raised venous pressures manifested as congested neck veins, haemodynamic compromise manifested as tachycardia and hypotension and also muffled heart

sounds.<sup>5</sup> Pericardiocentesis is performed to investigate moderate to large pericardial effusions and to gain urgent therapeutic benefits in cardiac tamponade. It is best practice to perform pericardiocentesis under ECHO and/or fluoroscopic guidance.<sup>4</sup>

Idiopathic pericardial effusion is described when it persists for three months but no cause is found despite active investigations. Small idiopathic pericardial effusions without haemodynamic compromise doesn't require follow up. While opposite is true for moderate to large sized effusions.<sup>4</sup>

## CONCLUSION

Pericardial effusion is generally secondary to systemic diseases and malignancies are commonest cause in adult population. It is investigated according to clinical presentation, epidemiological factors, size of pericardial effusion and blood investigations to look for inflammatory or infective causes.<sup>4</sup>

Typically, it would be expected late in the clinical course of lymphomas but in this patient it was the first manifestation of the disease. Considered early on to be idiopathic as there were no features to suggest a cause, interestingly patient developed DLBCL subsequently.

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