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Clinical Challenges Anne Marie C. Flaherty, MSN, RN, APNc • Associate Editor

Heartaches: Malignant Pericardial Effusions

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71-year-old woman named B.D. presented to the emergency department with a two-week history of fatigue, cough, and dyspnea. She complained that morning of a sudden worsening of respiratory symptoms, and a chest x-ray demonstrated an enlarged cardiac silhouette and left pleural effusion. Routine laboratory tests showed anemia, elevated liver enzymes, and renal insufficiency. A computed tomography (CT) scan of the thorax was ordered, revealing lung and liver lesions with a pericardial effusion. A follow-up 2D echocardiogram showed a pericardial effusion with evidence of cardiac tamponade. B.D. deteriorated rapidly in the emergency department. She had a decreased level of consciousness and was hypotensive with distended neck veins and muffled heart sounds. An electrocardiogram showed a rapid atrial fibrillation with electrical alternans. A echo-guided pericardiocentesis was performed at bedside. B.D. suffered cardiac arrest during the procedure but was successfully resuscitated. Two liters of fluid were drained from the pericardial sac. A surgical referral was requested for a pericardial window and an oncology referral was made because this was B.D.'s de novo presentation of metastatic disease.

A few weeks later, a 61-year-old man named S.T. presented to the chemotherapy clinic with a one-week history of increasing shortness of breath, an inability to lie flat, profound fatigue, and ankle swelling. S.T. had been receiving chemotherapy for a diagnosis of small cell lung cancer, and a CT scan showed shrinkage of his lung lesions but evidence of pleural and pericardial effusions. On examination, S.T. was tachycardic with a pulse of 150, a blood pressure of 90/60, and a pulsus paradoxus of 30 mm/Hg. Referral to a cardiologist and subsequent admission to the cardiac care unit resulted in pericardiocentesis (1 L of straw-colored fluid removed) and, ultimately, a pericardial window was created.

Definitions

A malignant pericardial effusion (MPE) is an accumulation of excess fluid in the pericardial sac that surrounds the heart and is associated with both solid tumor cancers such as lung or breast as well as hematologic malignancies such as leukemia (Cope, 2011). MPE is the most common cardiac complication and is a poor prognostic factor with survival usually less than six months (Nguyen, 2008).

Cardiac tamponade is a life-threatening complication of a pericardial effusion. It most often is caused by compression of the heart by fluid and results in inadequate filling of the ventricles, decreased cardiac output, and impaired cardiac function (Kaplow, 2011). Cardiac tamponade also can be caused by constrictive pericarditis as a result of radiation, either with acute inflammation or development of fibrous tissue as a late effect (Hoit, 2013).

Anatomy and Pathophysiology

Surrounding the heart are two distinct sacs—the endocardium and the pericardium. The endocardium consists of a layer of endothelial cells and an underlying layer of connective tissue. The pericardium is a two-layered sac (parietal and visceral layers) that contains the heart and great vessels that come from the heart. This sac contains a small amount of fluid (about 50 cc) that prevents friction and provides lubrication. Fluid is constantly moving in and out of this space. When the fluid increases, pressure and subsequent compression of the heart may occur. The amount of fluid in the sac is related to the degree of compression of the heart.

The sac itself can stretch and fill with almost 4 L of fluid. To maintain adequate cardiac output, the heart pumps faster and compensatory peripheral vasoconstriction occurs. Pericardial effusions can develop slowly over time or rapidly in a few hours or days. The faster the fluid collects, the more severe the distress. Although the volume is important, the speed of accumulation causes more problems because of the inability of the heart to adapt to the compression in a short period of time.

The literature shows that, at autopsy, about 20% of patients with cancer had metastatic disease in the pericardium (Newton, Hickey, & Marrs, 2009).

Diagnostic Tests

Many effusions are never detected, whereas others may be found via echocardiography, occasionally on chest xray, but most often on initial or interval extent of disease CT scans. Echocardiography has about a 96% diagnostic accuracy. Chest x-ray may highlight an increased pericardial silhouette with a water bottle-shape appearance on chest x-ray. Typically, electrocardiograms show only sinus tachycardia, but may demonstrate electrical alternans (an alteration in the amplitude of the QRS secondary to the mechanical swinging of the heart seen in large effusions) (Kaplow, 2011).

Culptic Cancers Associated With Pericardial Effusions

Lung and breast cancers, melanoma, and leukemias or lymphomas may be associated with pericardial effusions: lung and breast by direct extension or via the lymphatic route, and the blood-related malignancies metastasize and are present through hematogenous routes. A primary tumor rarely is found in the pericardium, but soft tissue sarcoma and mesothelioma have been diagnosed. Some therapies are associated with pericardial disease, such as cyclophosphamide, ifosfamide, all-trans retinoic acid, doxorubicin, and radiation to the thoracic region (Shanholtz, 2011).

Signs and Symptoms and Clinical Presentation

Detection of a pericardial effusion may not be easy, and many patients with a pericardial effusion have no symptoms (Halfdanarson, Hogan, & Moynihan, 2006). Accumulating fluid in the pericardial sac over a period of time enables it to stretch and accommodate the increasing volume. An acute and/or rapid increase in the volume is much more compelling and easier to discern. Figure 1 lists the signs and symptoms of pericardial effusion and cardiac tamponade.

The presence of hypotension, distended neck veins, and muffled heart sounds are known as Beck's Triad and should alert the healthcare provider to suspect cardiac tamponade. Patients with Beck's Triad are almost always anxious, restless, and in visible distress.

- With slow fluid accumulation: tachycardia, fatigue, dyspnea, hepatomegaly, abdominal distension, peripheral edema, and chest fullness
- With faster fluid accumulation: tachycardia, tachypnea, dyspnea, orthopnea, chest pain, and restlessness
- As pericardial effusion increases and progresses to cardiac tamponade: worsening tachypnea and cough, increased central venous pressure causing distended neck veins, pulsus paradoxus, hypotension, decreased or absent apical pulse, narrowed pulse pressure, confusion or dizziness, hoarseness, cough, hiccups, dysphagia, vasoconstriction, and cyanosis
- If no emergent intervention: Significant decrease in cardiac output leads to prolonged hypotension, resulting in organ failure and cardiac arrest.

Figure 1. Signs and Symptoms of Pericardial Effusion and Cardiac Tamponade

Note. Based on information from Cope, 2011; Kaplow, 2011.

Treatment

For a patient who is asymptomatic with compensatory hemodynamic mechanisms in place so that, except for mild tachycardia (pulse of about 100 beats per minute), adequate cardiac output occurs, then time is available to prepare for a pericardial window.

Cardiac tamponade is suspected if symptoms such as dyspnea or orthopnea, cough, retrosternal chest pain, anxiety, pulsus paradoxus (a drop in systolic blood pressure of greater than 10 mm/Hg during inspiration), hypotension, obvious cyanosis, and muffled heart sounds are present (Kaplow, 2011). Cardiac tamponade is an oncologic emergency and the goal of treatment is to remove fluid from the pericardium sac to relieve compression and prevent cardiovascular collapse. The level of intervention is dependent on the status of the patient, such as where they are in their illness trajectory, their age, comorbid conditions, the goal of treatment, and, most importantly, the patient's wishes. Preventing the reaccumulation of fluid also is an important treatment goal. Figure 2 lists the different interventions to manage pericardial effusion and cardiac tamponade. Pericardiocentesis will decompress the heart and immediately relieve symptoms and distress. It can be an initial treatment, particularly in patients with a poor performance status, and has a control rate of about 67% (Apodaca-Cruz et al., 2010).

The pericardial window or subxiphoid pericardiostomy is a surgical intervention that may be the initial procedure to treat a pericardial effusion. It also is indicated in recurrent effusions. This procedure has consistently demonstrated low morbidity and mortality rates and a recurrence rate of only about 4% (Nguyen, 2008).

Pharmacotherapeutics can play a role if the tumor is particularly chemotherapy sensitive (e.g., small cell lung cancer) and the clinician thinks the patient can tolerate treatment. Therapy should ensue in a timely fashion. The instillation of a sclerosing agent (e.g., doxycycline, bleomycin) may cause irritation and subsequent fibrosis to the pericardium and is reputed to work in about 50% of cases (Newton et al., 2009).

Nursing Interventions

Nursing interventions are many and are largely aimed at minimizing the

Balloon Pericardiotomy

- Via a percutaneous approach, a guide wire is threaded to the pericardium and then a balloon-dilating catheter is inserted and inflated to create a pericardial window.
 Less morbidity
- Less morbidity

Pericardectomy

- More extensive resection of the pericardium
- High morbidity and mortality—reserved for patients with low risk, good performance, and better prognosis
- Fewer complications with video-assisted thoracoscopic surgery approach versus thoracotomy
- Only definitive treatment for constrictive pericarditis

Pericardiocentesis

- Considered first-line therapy for immediate hemodynamic stabilization
- A needle is inserted into the pericardial sac to remove fluid under ultrasound guidance of a two-dimensional electrocardiogram or with electrocardiogram monitoring.
- A catheter can be inserted to continue to drain the fluid.

Pericardiocentesis With Sclerosing Agent

• For recurrent effusions, agents (i.e., doxycycline and tetracycline) are instilled after drainage to cause fibrosis of the pericardium, which prevents reaccumulation.

Subxiphoid Pericardiostomy (Pericardial Window)

- Small piece of anterior pericardium is removed.
- General anesthesia and stable cardiopulmonary status is required.

Systemic Treatment

- Palliative chemotherapy can be given to control effusions.
- Palliative radiotherapy is typically used with hematologic malignancies and breast cancer but can cause pericarditis.

Figure 2. Interventions for Pericardial Effusion and Cardiac Tamponade

Note. Based on information from Cope, 2011; Kaplow, 2011.

severity of the symptoms by helpful positioning (i.e., elevating the head of the bed to 90 degrees), application of oxygen, administration of IV fluid and medications to maintain cardiac output, managing pain, conserving the patient's energy, and dealing with other symptoms as they arise.

Assisting with a pericardiocentesis includes preparing the patient, monitoring hemodynamic and cardiac status,

Clinical Highlights

Malignant Pericardial Effusions

- Be alert to the possibility of malignant pericardial effusions (MPEs) in patients with subtle complaints such as fatigue, tachycardia, chest fullness, and anxiety. These patients will be unable to lie down and prefer sitting up and leaning forward.
- Patients most at risk include those with intrathoracic malignancies—breast and lung cancers, hematologic malignancies, history of thoracic radiation, and those currently anticoagulated.
- Severity of symptoms is strongly correlated with the rapidity of fluid accumulation. The faster the fluid accumulates, the more dramatic the cardiopulmonary decompensation.
- A 2D echocardiogram is the initial and most sensitive test to definitively diagnose MPE.
- Note incidental findings on computed tomography scans, such as small pericardial effusion, to identify a patient at risk.
- Although the patient may clinically give the appearance of congestive heart failure—with dyspnea, tachycardia, and distended neck veins—a pericardial effusion can lead to cardiac tamponade, causing compression of the heart and poor cardiac output. If hypotensive, these patients require vigorous fluid resuscitation until definitive therapy is initiated.

administrating medications and fluid, sending specimens for analysis, and observing for complications (Kaplow, 2011). Providing support and education to the anxious, distressed patient as well as his or her family are key to helping them understand the cause of the problems and plan for their resolution.

MPE with cardiac tamponade is an oncologic emergency and early recognition is key to the best outcome. As patients with cancer live longer and are afforded more treatment options for their diseases, the incidence and prevalence of MPEs are only likely to increase.

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