CLIINICAL VIGNETTE

Recurrent Pericarditis in a 57-Year-Old Woman

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A 57-year-old woman presented to the emergency department complaining of central sub-sternal chest discomfort. She described the pain as sharp and worse with coughing and rapid movement. She recalled that the pain came on suddenly, radiating into her neck and partially relieved by remaining completely still. She denied fevers, chills, cough, recent upper respiratory symptoms, nausea, diaphoresis or emesis. She denied any recent sick contacts, travel or recent new medication use. She had never been diagnosed with tuberculosis nor had any contacts. She had no personal or family history of coronary artery disease, autoimmune disease or malignancy. Her previous medical history was significant for very similar symptoms two years previously after a brief respiratory illness when she was diagnosed with viral pericarditis with a small pericardial effusion. She underwent a comprehensive workup to exclude autoimmune disease, neoplasia or drug related causes. She was told her heart condition was the result of a viral infection. Her symptoms resolved with non-steroidal anti inflammatory medications. On physical exam, her heart rate was eighty and regular, her blood pressure in her right arm was 134/77 and in her left arm was 130/74. She had no raised jugular venous distention; her carotid upstrokes were brisk bilaterally and of normal character and regular in rhythm. Her thyroid was not palpable and her apex beat non-displaced. Auscultation of her heart revealed a coarse biphasic scratchy murmur over the cardiac apex with the patient leaning forward at end exhalation. The murmur did not radiate. The remainder of her exam was unremarkable. Her twelve lead ECG revealed normal sinus rhythm at 78 beats per minute, normal intervals, without any ST or T wave changes. Her labs were notable for a white blood cell count (WBC) at 16 cells/mm^3 as well as an elevated C-reactive protein (CRP). Serial troponins were within normal range and the erythrocyte sedimentation rate (ESR) was normal. A chest x-ray was also unremarkable. Based on the history, physical exam and laboratory findings a diagnosis of recurrent pericarditis was made.

Recurrent or relapsing pericarditis occurs in up to 30% of patients but ranges from 15-32% by various reports[1, 2]. Patients usually present with typical pleuritic chest pain accompanied by a precordial friction rub, leukocytosis, fever and elevated inflammatory markers. The condition is defined as: "A syndrome in which acute pericarditis recurs after the agent inciting the original acute attack has disappeared or has ceased to be active." The predominant feature of recurrent pericarditis is pleuritic chest pain, and other features of acute pericarditis may be absent[2]. Symptomatic recurrence can be at any point following the resolution of the initial acute pericarditis symptoms, but usually occurs within weeks to months. In those in whom symptoms never resolve after the initial episode, the term "incessant pericarditis" is used. The criteria for diagnosis used in the landmark colchicine for recurrent pericarditis trial (CORE) [3] are widely used as the standard for making a diagnosis [Box 1].

[Box 1] The COlchicine for REcurrent Pericarditis (CORE) Trial Criteria[3]

Criteria for Recurrence (Need both)	1. A documented prior attack of acute pericarditis of idiopathic, viral, or autoimmune origin 2. Recurrent Pleuritic Chest Pain
Additional Criteria	Evidence of either recurrence or continued activity of pericarditis

One or more signs of inflammation:	• Fever
	• Pericardial friction rub
	• Typical ECG changes:
	  o Diffuse ST segment elevation
	  o PR segment depression
	• Echocardiographic evidence of pericardial effusion
	• Cardiac MRI evidence of inflammation of the pericardium
	• An elevation in (1 or more):
	  o WCC
	  o ESR
	  o CRP

Most cases of relapsing pericarditis are caused by recurrent autoimmune processes. The evidence to support this includes the prolonged latent period, the presence of antibodies and a response to glucocorticosteroids and other immunosuppressive treatments[4]. Proving an infectious cause for the recurrence is difficult and invasive (e.g. pericardiocentesis if effusion is present). One study used pericardioscopy, multiple epicardial biopsies, and polymerase chain reaction (PCR) and found a 23 percent higher incidence of infection or reinfection, mostly of a viral cause[4]. A 2004 European Society of Cardiology guideline introduced the term *autoreactive pericarditis* which refers to a diagnosed case of recurrent pericarditis but where no evidence of infection, autoimmune disease or malignancy is found[5]. This condition is widely felt to be a local rather than systemic immune response.

The first line treatment for most patients with recurrent pericarditis (viral, autoimmune, idiopathic) is colchicine combined with a non-steroidal antiinflammatory drug (NSAID) (Grade 1A) [4]. Indomethacin is preferred to ibuprofen and should be used for at least one month. Treatment response is gauged by symptom resolution and decline of serum inflammatory markers. Glucocorticoids may be considered in patients with diagnosed systemic autoimmune disease, allergy or intolerance to NSAIDS or who fail combination therapy with colchicine and NSAIDS[5]. Glucocorticoid dosing is based on consensus opinion rather than controlled trial data and starts at a prednisone dose of 0.25 to 0.50 mg/kg/day followed by a slow taper[6]. Intrapericardial steroid therapy, prolonged immunosuppression, and pericardiectomy are therapeutic options for that minority of patients with continued recurrences or whose symptoms persist despite standard therapy. Patients with idiopathic recurrent pericarditis usually have a good prognosis[7].

Our patient received oral colchicine at 1 mg on the first day, followed by 0.5 mg twice daily for two months. Initially, she overlapped colchicine treatment with ibuprofen 600 mg three times a day for 2 weeks and was noted to be symptom free at 8 months follow-up.

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