

# Inflammation and Rupture of a Congenital Pericardial Cyst

Manifesting Itself as an Acute Chest Pain Syndrome

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We present the case of a 63-year-old woman with a remote history of supraventricular tachycardia and hyperlipidemia, who presented with recurrent episodes of acute-onset chest pain. An electrocardiogram showed no evidence of acute coronary syndrome. A chest radiograph revealed a prominent right-sided heart border. A suspected congenital pericardial cyst was identified on a computed tomographic chest scan, and stranding was noted around the cyst. The patient was treated with nonsteroidal anti-inflammatory drugs, and the pain initially abated. Another flare-up was treated similarly. Cardiac magnetic resonance imaging was then performed after symptoms had resolved, and no evidence of the cyst was seen. The suspected cause of the patient's chest pain was acute inflammation of a congenital pericardial cyst with subsequent rupture and resolution of symptoms. (*Tex Heart Inst J* 2016;43(6):537-40)

**P**ericardial cysts are a rare congenital anomaly, and most patients who have them are asymptomatic. When symptoms do occur, they are generally the result of an anatomic, space-occupying effect that displaces surrounding structures. Pericardial cysts are most often diagnosed as an incidental finding. Here, we discuss the case of a patient who developed a recurrent acute chest-pain syndrome related to inflammation, and the subsequent rupture of a congenital pericardial cyst.

## Case Report

On 23 June, a 63-year-old woman with a history of supraventricular tachycardia and hyperlipidemia presented with acute-onset, severe chest pain. She had been bothered by what she suspected was an upper respiratory tract infection in the 3 weeks preceding her initial presentation. During this time, her main symptom had been a nonproductive cough. She had taken an airplane flight from Los Angeles to Houston on June 21. Then, in the early morning on June 23, she developed severe, sharp, stabbing, unrelenting chest pain. There was no improvement or worsening with position changes or breathing. She felt that she could not catch her breath.

She presented at her cardiologist's office. She was afebrile with otherwise normal vital signs, and the results of a physical examination were unrevealing. An electrocardiogram showed sinus rhythm with no ST-segment or T-wave changes to suggest acute coronary syndrome (Fig. 1). Laboratory results included a creatinine level of 0.93 mg/dL; sodium, 136 mEq/L; hemoglobin, 11.5 g/dL; white blood cell (WBC) count, 6,700/ $\mu$ L, with normal differential; and platelets, 234,000/ $\text{mm}^3$ . Her liver-function tests were all within normal limits. Her C-reactive protein (CRP) was markedly elevated at 105 mg/L. A chest radiograph showed a mildly prominent right-sided heart border, and the lung fields were clear (Fig. 2).

The patient was referred to a pulmonologist. A computed tomographic (CT) chest scan showed no evidence of pulmonary embolism. The aortic root was mildly dilated at 4.1 cm, but there was no evidence of dissection. The CT angiogram did reveal a suspected congenital pericardial cyst (size, 2.3  $\times$  4.2 cm) along the right-sided heart border (the patient was known to have a pericardial cyst, an incidental finding on a CT scan in 2008). Because of the diagnostic study's failure to produce a definitive diagnosis, she was managed conservatively with naproxen and pantoprazole, and her pain subsided over the next few days.

**Key words:** Cysts/complications/pathology; pericardial cyst, congenital; pericardium/abnormalities; rupture, spontaneous

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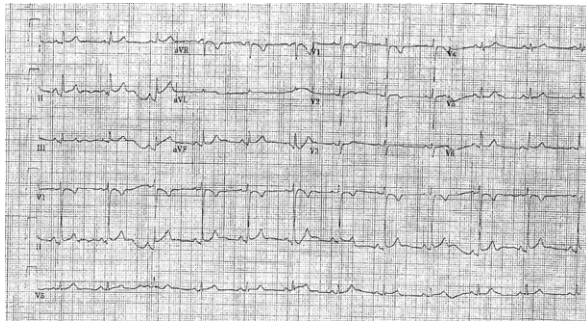
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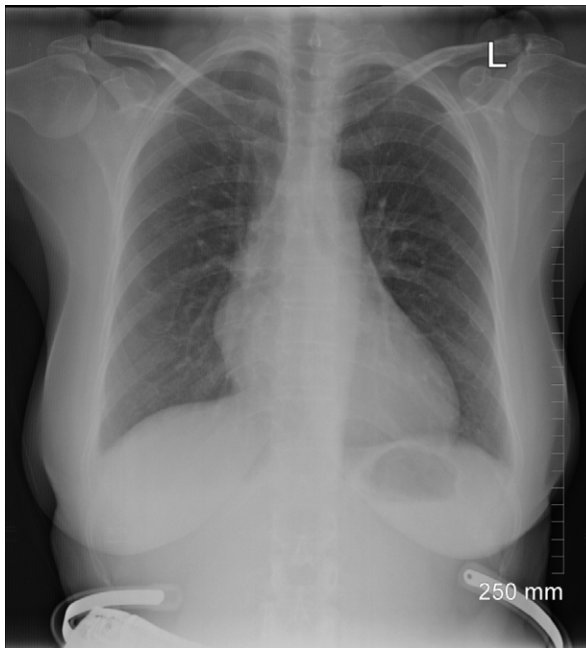
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In the early morning hours on 13 July, the patient had acute and recurrent episodes of sharp, crushing chest pain. She felt extremely weak and could barely get out of bed. She managed to check her blood pressure at home and the systolic pressure was over 170 mmHg. She felt dizzy upon standing but never lost consciousness. She reported no shortness of breath or cough with this episode.

The patient went to work, then subsequently sought medical attention. Physical examination results and vital signs were again unremarkable. Laboratory results included a CRP level of 53 mg/L; an erythrocyte sedimentation rate of 16 mm/hr; a WBC count of 8,000/ $\mu$ L, with a normal differential; hemoglobin, 11.7 g/dL; and platelets, 194,000/ $\text{mm}^3$ . A chest radiograph showed a more enlarged and irregular right-sided heart border

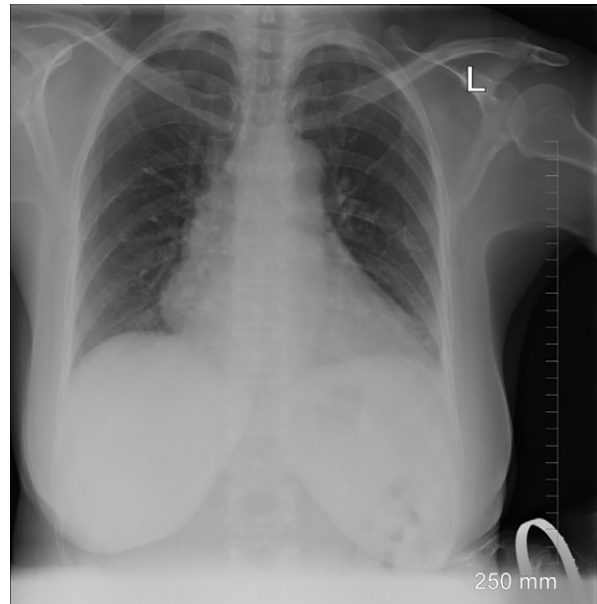


**Fig. 1** Electrocardiogram on 23 June shows no evidence of acute coronary syndrome.

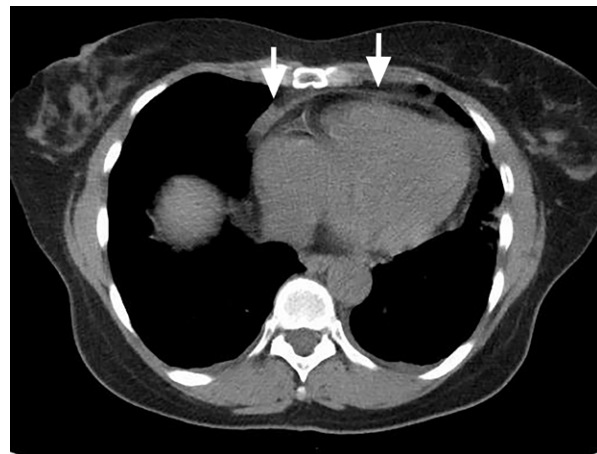


**Fig. 2** Chest radiograph on 23 June shows a mildly prominent right-sided heart border and clear lung fields.

than did her prior radiograph (Fig. 3). A repeat CT chest scan showed 2-mm-thick pericardium (within the reference range), but there was notable stranding along the pericardium extending to the cyst, which was new in comparison with the previous study (Figs. 4 and 5). The pericardial cyst was again seen (size, 2.5  $\times$  4.9 cm), and no hemorrhagic component was noted. There was concern that the pericardial cyst and pericardium were inflamed and contributing to her symptoms. Because the images and laboratory reports suggested pericardial inflammation, we prescribed 800 mg of ibuprofen, 3 times daily. We also prescribed colchicine, but the patient declined to take it.



**Fig. 3** Chest radiograph on 13 July shows a prominently enlarged and irregular right-sided heart border.

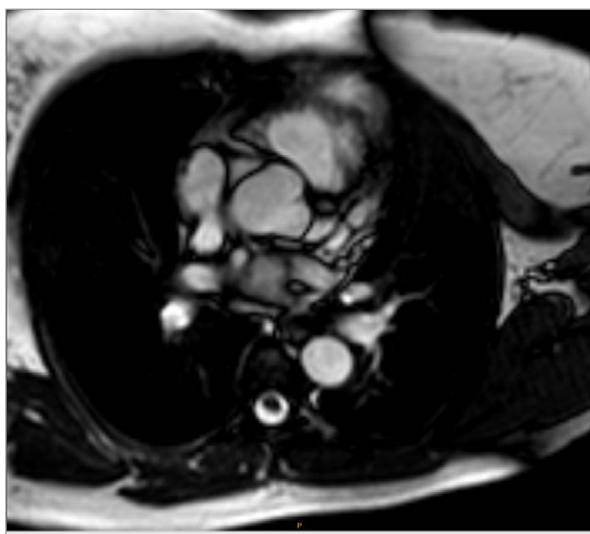


**Fig. 4** Computed tomographic scan (non-contrast) on 13 July shows stranding and thickening (arrows), suggesting an inflamed pericardium.

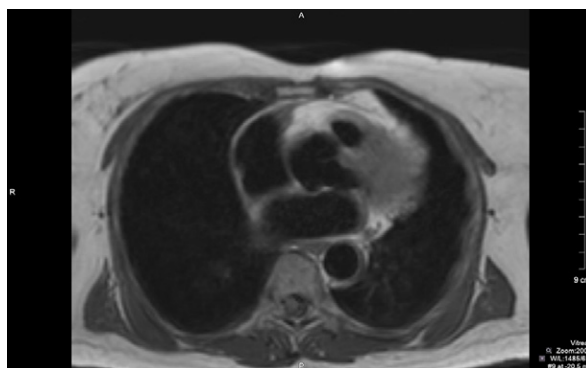
The patient's chest pain resolved over the next several days; she was left with a sensation of "chest heaviness" that persisted for several more weeks, but resolved by early August. Cardiac magnetic resonance images on 13 August showed no evidence of a pericardial cyst or stranding, despite our using multiple imaging techniques (Figs. 6 and 7). Inflammatory markers were repeated and were within normal limits on follow-up. In outpatient monitoring over the ensuing year, the patient has reported no further recurrences.



**Fig. 5** Computed tomographic scan on 13 July shows a pericardial cyst (PC).



**Fig. 6** Cardiac magnetic resonance image (cine gradient echo) on 13 August shows no evidence of a pericardial cyst after resolution of chest discomfort.



**Fig. 7** Cardiac magnetic resonance image (T1 spin echo) on 13 August shows no evidence of a pericardial cyst.

## Discussion

We attribute this patient's acute chest pain to an inflamed congenital pericardial cyst, with subsequent rupture. This conclusion is supported by her patently elevated inflammatory markers with CT images revelatory of stranding along the pericardium and into the cyst, and suggestive of an acute process. As symptoms resolved, her inflammatory markers normalized. Once symptoms had completely resolved, repeat cross-sectional images showed that the cyst was no longer present, most likely indicative of cystic rupture.

Pericardial cysts are a rare and generally an incidental finding. The estimated prevalence is 1 in 100,000 people.<sup>1,2</sup> They are most often seen at the cardiophrenic angles. Pericardial cysts are thought to be congenital in origin and related to persistence of a ventral recess in the pericardial celom.<sup>3</sup>

Most congenital pericardial cysts cause no symptoms. The authors of a large case series of 82 patients affected by pericardial cyst estimated that about 40% of patients have related symptoms.<sup>4</sup> Other (and smaller) case series indicate higher rates of symptomatic patients—but given the small numbers in these series, selection bias most likely contributes to the higher rates of symptomatic patients.<sup>5-8</sup> The symptoms typically are chest pain or discomfort, dyspnea, cough, and arrhythmias.

Sequelae can arise from pericardial cysts. These are generally hemodynamic and obstructive problems related to the cyst size and displacement of the heart or great vessels. Reported sequelae include cardiac tamponade, right ventricular outflow tract obstruction, sudden cardiac death, syncope, and congestive heart failure. Case reports have detailed the destructive nature of some cysts, including cystic erosion into the superior vena cava and right ventricular free wall that results in hemorrhage.<sup>9,10</sup> Congenital pericardial cystic rupture is reported infrequently,<sup>11,12</sup> but multiple reports document catastrophic rupture of pericardial and myocardial hydatid cysts, which appear to have a more malignant

course than do congenital pericardial cysts.<sup>13-15</sup> By consensus, the treatment for cardiac hydatid cysts—which are most often intramyocardial but can also be confined to the pericardial space—is a combination of surgical resection (for all patients who can tolerate it) and albendazole.<sup>16,17</sup>

Management strategies have been proposed for congenital pericardial cysts. Often, either aspiration or surgical resection is indicated in patients who are thought to be symptomatic because of the pericardial cyst. Conservative management and observation are indicated in patients who are asymptomatic and do not manifest cystic enlargement upon serial imaging.<sup>8</sup> If there is uncertainty about the cause of the cystic structure (for example, malignancy), then sampling of the tissue could be indicated.

### Acknowledgment

We appreciate the help of M.C. Davault in the preparation of this manuscript.

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