

# Pericardial Fat Necrosis

## A Review and Update

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*A previously healthy middle-aged person presents with excruciating left-sided chest pain of 6 hours' duration. The pain has come on abruptly, without warning, and is located in the lower part of the chest anteriorly. It radiates to the neck and left shoulder and worsens on deep inspiration. The patient appears seriously ill, with tachypnea, tachycardia, and diaphoresis. Otherwise, the physical examination is unremarkable. The electrocardiogram shows sinus tachycardia. Results of conventional blood studies and the chest radiograph are within normal limits. Three days later, a follow-up chest radiograph shows a 3.5 × 4-cm mass adjacent to the left side of the heart near the diaphragm.*

**T**his hypothetical case illustrates the typical presentation of a rare but important benign disease—pericardial fat necrosis (PFN). At least 23 cases of PFN have been documented in the English-language medical literature since Jackson, Clagett, and McDonald described the condition in 1957.<sup>1-17</sup> Textbooks of internal medicine<sup>18-22</sup> and cardiology<sup>23-25</sup> offer nothing on this ailment, and only 1<sup>26</sup> of 3<sup>26-28</sup> books devoted solely to the pericardium mentions it. As a result, PFN remains little known and poorly appreciated. Yet, early in its course, PFN characteristically is mistaken for a serious disease, particularly myocardial infarction<sup>1-3,10</sup> or pulmonary embolism.<sup>1,8,10,13</sup> Later, it uniformly mimics a pericardial cyst or a pericardial or pulmonary neoplasm. Misdiagnosis and mismanagement are inevitable, therefore, if one is not familiar with this unique clinicoradiologic entity. Hence, this review.

### Clinical Features

Pericardial fat necrosis classically strikes suddenly, without warning. All of the victims reported to date—15 men and 8 women—were in good health at the time PFN began. They ranged in age from 23 to 67 years (mean age, 49.2 yr). Five of the men<sup>1-3,6</sup> and 2 of the women<sup>11,12</sup> were white and 1 of the women was black.<sup>13</sup> Race was not recorded for the remaining 15 patients.

Severe chest pain, typically pleuritic, is the initial manifestation. It was left-sided in 15 patients,<sup>1-3,5,7,9-11,13,14,16</sup> right-sided in 5,<sup>1,4,8,10,12</sup> present but not further described in 1,<sup>16</sup> absent in 1,<sup>17</sup> and not mentioned in 1.<sup>15</sup> The pain is located anteriorly near the diaphragm and radiates at times to the neck, shoulder, upper arm, axilla, or back. It lasts several days to a week or so, but can recur with less intensity for up to a year.<sup>1,10</sup> Fever and cough are not features of PFN.

If examined soon after onset of the chest pain, the patient is dyspneic, with tachypnea, tachycardia, and diaphoresis. One patient was hypotensive at admission,<sup>3</sup> 2 had a pericardial friction rub,<sup>3,7</sup> and 3 others had marked tenderness to palpation over or near the precordium.<sup>5,6,10</sup> By contrast, if several days or more elapse before the patient comes under observation, the physical examination usually gives normal findings.

**Electrocardiographic Observations.** The electrocardiogram characteristically is normal. Occasionally, it shows tachycardia, nonspecific ST- or T-wave changes suggesting ischemia, or findings consistent with resolving pericarditis.<sup>8,13,14</sup> In 1 case, it showed right bundle branch block.<sup>11</sup>

### Imaging Results

Chest radiographs obtained during the first few days of the illness may show no abnormality. Thereafter, a mass invariably appears in or near the cardiophrenic angle on the side of the chest pain. The mass is always located anteriorly and is almost al-

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ways contiguous with the cardiac silhouette. In 1 case, however, it extended between the lingula and left lower lobe<sup>10</sup>; in another, it overlay the left hemidiaphragm in the area of the interlobar fissure<sup>1</sup>; and in another, it was distinctly separate from the heart.<sup>7</sup> Finding such a mass on the chest radiograph always raised concern for a pericardial cyst or a pericardial or pulmonary neoplasm.

Computed tomography (CT) helps determine the nature and exact location of the chest mass.<sup>12-16,17</sup> From density measurements, one can infer that the mass consists of fat; but CT cannot always distinguish between a benign and malignant fatty tumor, especially when stranding is present within the fat.<sup>13,16</sup> In such cases, operative intervention might still be indicated to establish the precise diagnosis.

Magnetic resonance imaging performed in 2 patients confirmed the CT findings of fat with dense stranding.<sup>14,16</sup>

Gallium-67 scintigraphy in a patient with unexplained hemoptysis incidentally showed increased gallium uptake in the pericardial fat.<sup>15</sup> Subsequent biopsy of the pericardial fat showed necrosis but no evidence of malignancy.

### Miscellaneous Studies

Pleural effusion occurred in 4 patients. Thoracentesis yielded only a small amount of bloody fluid in 1 case,<sup>1</sup> “did not demonstrate any evidence of malignancy and the bacterial culture was negative” in another case,<sup>14</sup> and was not mentioned in the third case.<sup>15</sup> In the fourth case, thoracentesis disclosed a pH of 7.58; erythrocytes, 13,600/mm<sup>3</sup>; leukocytes, 5,000/mm<sup>3</sup> (64% segmented neutrophils, 13% lymphocytes, and 24% monocytes); glucose, 139 mg/dL; protein, 4.6 g/dL; and lactate dehydrogenase, 167 U/mL.<sup>13</sup>

Transthoracic and transesophageal echocardiography performed in 1 patient<sup>17</sup> demonstrated a 5-cm, ovoid, solid pericardial mass compressing the right side of the heart.

Other investigations that were noncontributory included the total and differential leukocyte counts, erythrocyte sedimentation rate, serum amylase and electrolytes, cardiac enzymes, barium swallow and barium enema, complete metastatic workup, arterial blood gas determination, ventilation/perfusion lung scan, bronchography, bronchoscopy with cytologic washings, pulmonary function tests, and pulmonary angiography.<sup>4,7,8,12,13,17</sup>

### Diagnosis

In 22 of the 23 cases, histologic examination of biopsied or (most often) resected tissue proved PFN. The sole exception was a case reported 5 years ago, in which the clinical, radiographic, and CT findings prompted a tentative diagnosis of PFN.<sup>16</sup> With symptomatic treatment, the chest pain resolved at 1 week, and the chest

radiograph 2 months later showed that the paracardiac density had disappeared. A follow-up chest CT at 2 months also showed a marked decrease in size and thickness of the pericardial lesion.

### Surgical and Histologic Findings

After nondiagnostic workups, sometimes extensive, 21 of the 23 patients underwent exploratory thoracotomy. At operation, the typical finding was an inflammatory mass involving the parietal pericardial fat pad; masses varied from 1.5 cm in size<sup>9</sup> to 10 × 7.5 × 3 cm in size.<sup>3</sup> The pathologic features bore close resemblance to those of infarcted epiploic appendices and to fat necrosis in the breast.<sup>8</sup> Lesions removed early in the clinical course showed a central focus of necrotic fat cells encompassed by macrophages with intense neutrophilic infiltration.<sup>12</sup> Specimens removed later in the clinical course showed considerable fibrosis as well. In 1 case, calcifications were also present.<sup>17</sup> Resection of the diseased tissue effected a cure in every case, with follow-ups for as long as 19 years.<sup>10</sup>

### Pathogenesis

The cause of PFN remains speculative. In 2 patients, the mass was attached to the heart by a pedicle, acute torsion of which might have triggered the necrosis.<sup>15</sup> A pre-existing structural abnormality of the adipose tissue, such as lipoma or hamartoma, might make the tissue vulnerable to the trauma of a beating heart and moving diaphragm.<sup>4,6,17</sup> Two cases lend credence to this possibility: in 1, lipomatosis abutted the right atrium<sup>17</sup>; in the other, the resected specimen was histologically consistent with a lipoma showing fat necrosis.<sup>4</sup> In the circumstance of extreme lifting efforts before<sup>6</sup> or during<sup>3</sup> onset of the chest pain, rapid changes in intravascular pressure associated with the Valsalva maneuver might cause hemorrhage into the loosely supported adipose tissue of the pericardium. The hemorrhage might, in turn, initiate the necrosis.<sup>6</sup> Although the original report on PFN listed obesity as a probable prerequisite for the disease,<sup>1</sup> only 8 of the 20 patients in subsequent reports were said to be obese.<sup>8-11,14</sup> Evidence of recent or concomitant infection, acute pancreatitis, or any other disease has been absent in every case.

### Comment

Pericardial fat necrosis is unique in that no other known disease causes sudden, excruciating, low anterior chest pain—pleuritic in type and without fever or cough—followed in a few days by a rapidly developing mass in or near the cardiophrenic angle. That sequence of events dictates the differential diagnosis—cardiopulmonary emergency early, neoplasm later. The chest pain eventually subsides, but the mass tends to persist with little or no change.

One final point: In 1958, a year after the initial description of PFN, Kasserman<sup>2</sup> suggested that excision of the necrotic inflamed mass “may not prove to be the ideal therapeutic approach if more is learned about this disease.” His prophecy became a reality in 2005 when Pineda and colleagues<sup>16</sup> reported the first and only case to date in which a tentative diagnosis of PFN led to non-surgical management and disappearance of the paracardiac mass on the chest radiograph within 2 months.

### Take-Home Message

Given the unique clinicoradiologic picture and benignity of PFN, coupled with CT’s ability to show that the chest mass has the density of fat, a clinical diagnosis and symptomatic care should suffice in most instances. Only when a serious diagnostic question remains or the patient suffers intractable pain should thoracotomy be necessary.

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