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The Fortress Becomes a Prison: Calcified Constrictive Pericarditis

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PRESENTATION

The pericardium, normally so useful in defending the heart from outside invaders, can perform more like a cage when its flexibility is compromised. We present the case of a 50-year-old man who was admitted to the nephrology unit when severe chronic hypotension led to poor hemodynamic tolerance during hemodialysis. He had a 24-year history of kidney disease of unknown etiology. At debut, he presented with pericardial effusion that required urgent initiation of hemodialysis. In 1993, he underwent a kidney transplant. Chronic graft dysfunction evolved, and in 2010, he started hemodialysis again. His medical history included chronic liver damage secondary to venous-occlusive disease induced by azathioprine and mesenteric thrombosis managed with oral anticoagulation. In 2011, he underwent subtotal parathyroidectomy for hyperparathyroidism.

ASSESSMENT

Upon evaluation, the patient had severe dyspnea, permanent jugular distension, severe hypotension, and slow capillary refill. A chest radiograph showed a high-density image outlining the pericardium (**Figure 1**). Transthoracic echocardiography revealed a thickened pericardium (6 mm) with normal systolic function and diastolic dysfunction. Computed tomography disclosed diffuse calcification and thickening of the pericardium (**Figure 2A**).

DIAGNOSIS

The patient was diagnosed with calcified constrictive pericarditis. Vascular calcifications are common among those who have end-stage renal disease. These calcifications have been associated with other factors, such as hyperphosphatemia, hyperparathyroidism, oral anticoagulation, and chronic inflammation. The main sites of vascular calcification in patients with end-stage renal disease include the medial layer of the arteries and the heart valves. However, calcified pericardium is rare in this population.

Pericarditis is frequently an idiopathic disease, but it can also occur after cardiotomy, acute myocardial infarction, radiation, tuberculosis, uremic syndrome, and systemic inflammatory disorders.[1] Pericardial inflammation in end-stage renal disease is a serious disorder, typically secondary to acute uremic syndrome or dialysis-related disease. Chronic constrictive pericarditis is unusual, and calcified constrictive pericarditis, in which the heart is encased within a rigid calcified pericardium, is even more exceptional.[2,3]

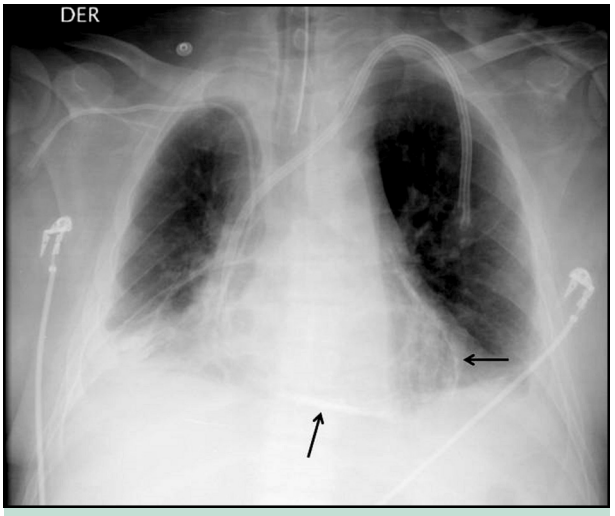

Figure 1 A chest radiograph showed a high-density image outlining the pericardium (arrows).

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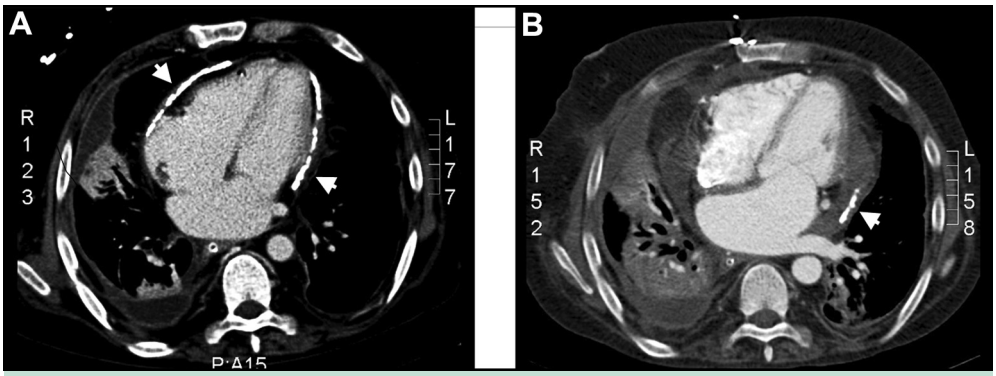

Figure 2 (**A**) Computed tomography identified diffuse calcification and thickening of the pericardium (arrowheads). (**B**) Pericardial tissue (arrowhead) remained after pericardiectomy.

Figure 3 This sample of pericardium was removed during surgery.

MANAGEMENT

Pericardiectomy is the treatment of choice for improving cardiac hemodynamics in patients with constrictive pericarditis. Our patient underwent this procedure because he had severe hemodynamic compromise from calcified constrictive pericarditis (**Figures 2B and 3**).[2,4] A pericardial biopsy showed severely calcified pericardium with no evidence of inflammatory activity (**Figure 4**). Ziehl-Neelsen staining was negative. After surgery, he remained hospitalized in the intensive care unit for a prolonged period. Nonetheless, he died after multiple organ failure.

The extreme degree of calcification identified in this patient is a rare condition. It can be explained by his long history of chronic kidney disease, uremic pericarditis at the initial diagnosis of end-stage renal disease, severe hyperparathyroidism, chronic hyperphosphatemia, and use of oral anticoagulation. His disease was so severe that his pericardium was transformed into a rigid cage, ultimately causing his death.

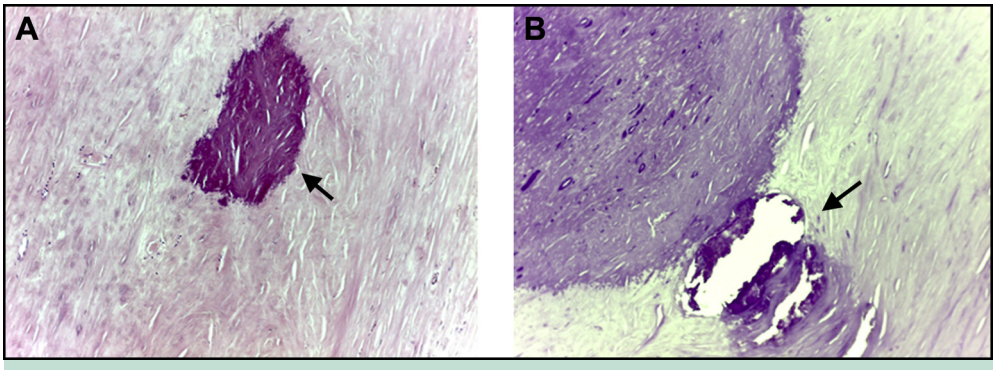

Figure 4 (**A**) Tissue was treated with nitric acid, 5%. Fibrous and hypocellular tissues with few capillary vessels are visible in this photograph. The darker area (arrow) correlates with the focus of calcification. (**B**) Here, the tissue has not been treated with nitric acid, 5%. This image shows foci of calcification (arrow) corresponding to a site of tissue disruption caused by the hard consistency of the calcification (hematoxylin and eosin, 40×).

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