



Letter to the Editor

## Constrictive pericarditis in Erdheim–Chester disease: An integrated echocardiographic and magnetic resonance approach



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Erdheim–Chester disease (ECD) is a rare non-Langerhans histiocytosis affecting middle-aged adults. The disease is named after the cardiology fellow (William Chester) and his mentor (Jakob Erdheim) who reported the first two cases in 1930 in Vienna as ‘lipoid granulomatosis’ [1]. Its etiology is unknown, and it is characterized by the proliferation of lipid-containing foamy histiocytes infiltrating bones and potentially every organs. Histiocytic infiltration leads to xanthogranulomatous infiltrates of multiple organ systems [2]. Symptoms vary on the basis of tissue involvement, most patients have bone pain however around half complain extraskeletal manifestations including exophthalmos, xanthelasma, interstitial lung disease, retroperitoneal “fibrosis” with perirenal or ureteral obstruction, renal failure, central nervous system and cardiovascular involvement. Most common cardiac manifestation is periaortic fibrosis with a “coated aorta” aspect, and pericardial and myocardial infiltration, intracardiac tumors, and valvular heart disease are less frequent clinical conditions [3,4]. We describe the only known case of histiocytosis infiltration involving pericardial tissue with consequent constrictive pericarditis in a middle aged patient with peritoneal infiltration as first manifestation.

A 46-year old man was previously hospitalized (2 years before) for global bone pain associated to a progressive renal dysfunction due to kidney infiltration and retroperitoneal infiltration secondary to primitive diseases. During the first admission bone and kidney biopsy revealed ECD. Therefore for better disease evaluation an abdominal

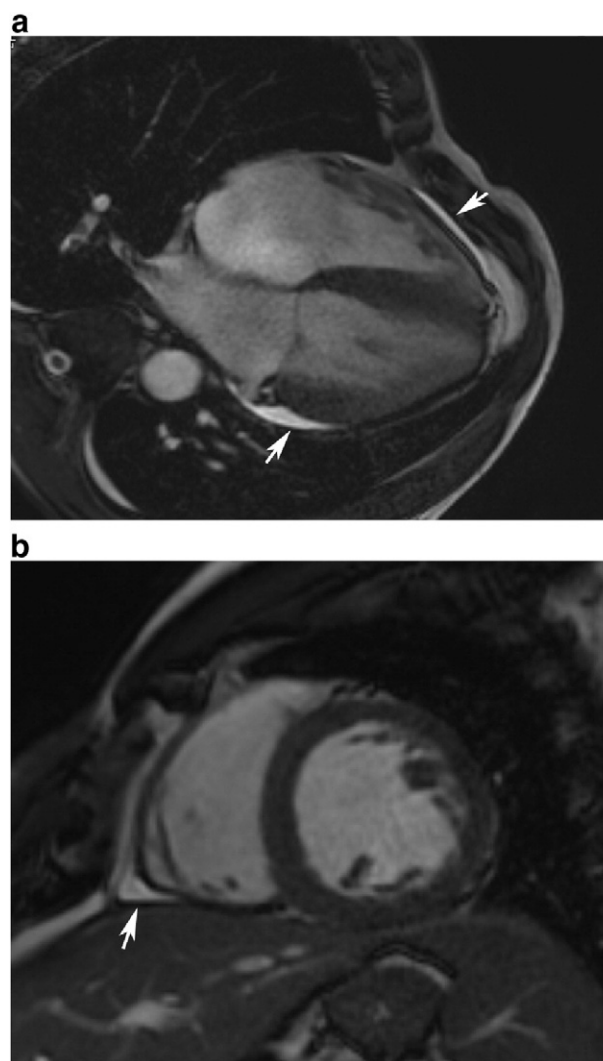


Fig. 1. Steady state free precession four chamber view (a) and short axis view (b) showing a mild pericardial effusion. The thickness of pericardial leaflets appears normal.

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and chest Computed Tomography (CT) scan was made. Cardiac magnetic resonance (CMR) was also performed and it revealed a mild pericardial effusion without tamponade evidence or any myocardial tissue infiltration (Fig. 1a and b). During the second admission, the patient was referred for shortness of breath, low blood pressure value, general asthenia and bilateral ankle swelling. Physical examination showed increase of heart rate (105 beats for minute), padded cardiac sounds, and mild jugular distention with venous pressure measurement of 12 cm H<sub>2</sub>O. Chest examination did not evidence rales but chest radiography revealed a large cardiac silhouette. ECG showed increased heart rate with a specific low voltage on precordial derivations. Transthoracic echocardiography showed left ventricular hypertrophy with septal and posterior wall thickness, good systolic function, reduced right sizes with decreased longitudinal function and enlarged inferior vena cava size

associated with reduced inspiration movement. Left atrial size and aortic root were into the limit. Pericardial evaluation pointed out an increased parietal leaflet and brightness with circumferential material deposition. Transmitral pulsed Doppler showed a clear restrictive filling pattern (Fig. 2a and b). Due to echocardiographic findings, the patient was submitted to a new CMR examination confirming the presence of extensive infiltrating tissue in the pericardium (Fig. 3a and b). To confirm pericardial involvement and restrictive pericarditis pericardiocentesis with bioptic drawing was done: histological specimen confirmed the presence of fibrotic material associated to abundance of foamy macrophage and histiocytes positive for CD68 but not for S-100 protein or CD1a (Fig. 4a and b). Thus, on the basis of echocardiographic and CMR examinations and histological findings, the diagnosis of a restrictive process affecting the pericardium was done.

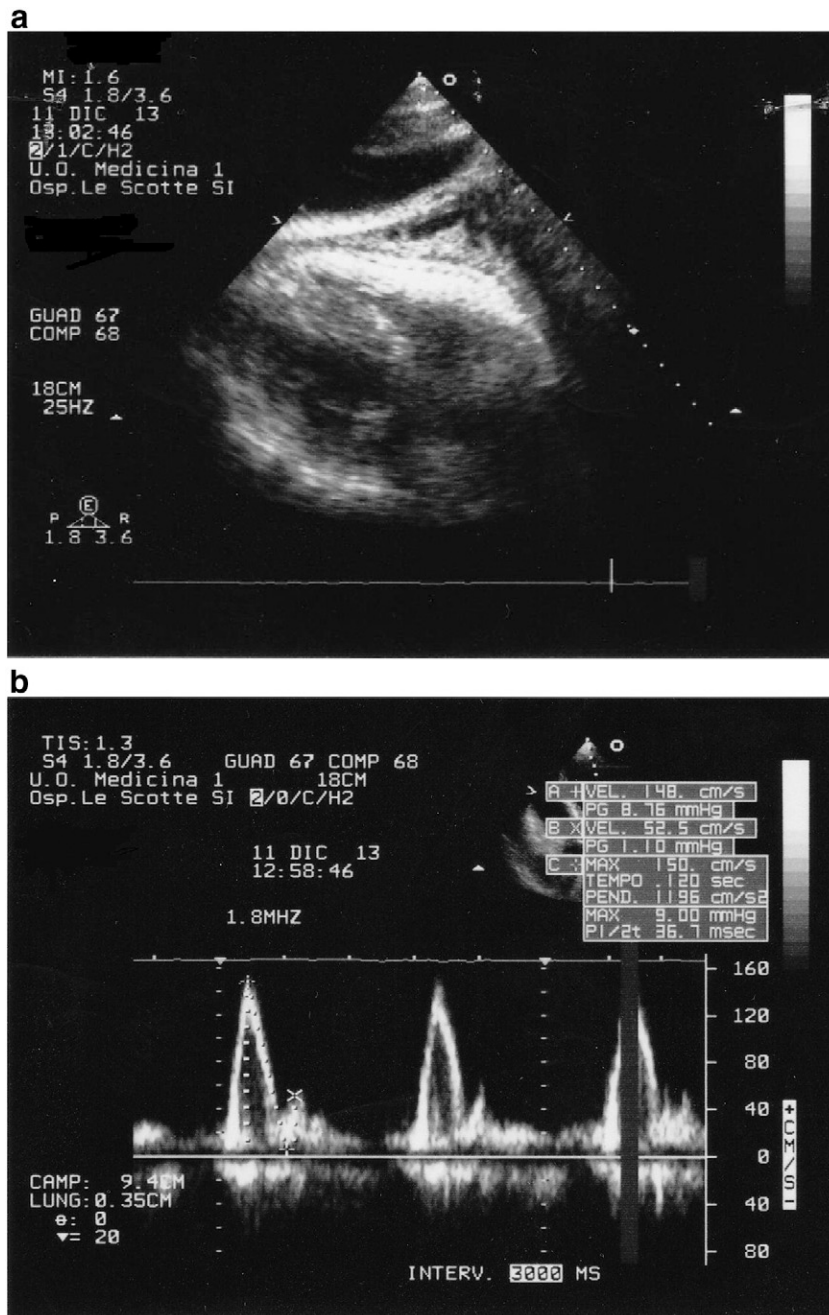


Fig. 2. Pericardial thickness associated with fibrinic material deposition and adherences between internal and external leaflets (a); pattern Doppler of transmitral flow demonstrating a classical “restrictive filling pattern” with increased E/A ratio and reduced deceleration timing (b).

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