

Image in cardiology

# Erdheim-Chester Syndrome With Periaortitis



## Síndrome de Erdheim-Chester con periaortitis

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Figure 1.

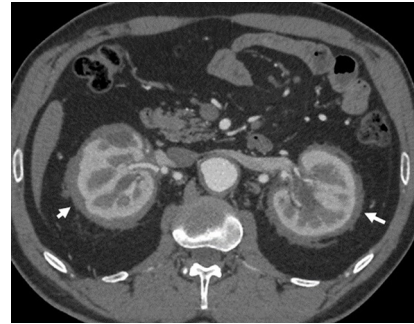


Figure 2.

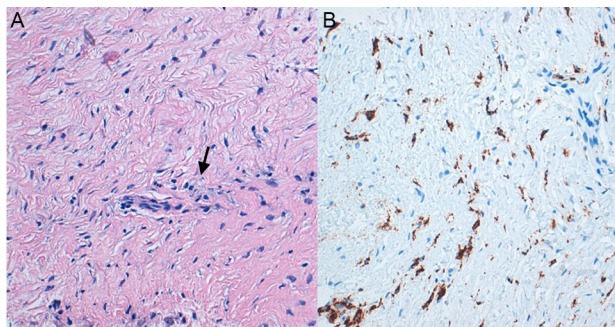


Figure 3.

Aortitis and periaortitis encompass a number of diseases, whose common feature is inflammation of the aortic wall. Their clinical presentation is variable, with nonspecific symptoms such as abdominal pain, fever, and weight loss. For this reason, imaging techniques are of great value.

The patient was a 47-year-old man with suspected acute aortic syndrome, in whom computed tomography angiography revealed circumferential thickening with thoracoabdominal periaortic soft tissue density, irregularity of the aortic lumen (Figure 1A, arrows), and ostial stenosis of the left subclavian artery, celiac trunk, and superior mesenteric artery (Figure 1B, arrows). The kidneys showed bilateral, symmetrical infiltration of the perirenal tissue with dilation of the renal collecting system (Figure 2, arrows). Perirenal biopsy showed extensive fibrosis with foamy macrophages (Figure 3A, arrow) and positive staining for CD68 (Figure 3B). The findings were compatible with Erdheim-Chester disease and treatment with pegylated interferon alpha was begun.

This disease is a rare form of nonhereditary histiocytosis that invariably affects the long bones. Retroperitoneal, cardiovascular, or nervous system involvement is detected in 50% of affected individuals. The disease is characterized by infiltration of the connective and perivascular tissue by foamy histiocytes that stain positively for CD68, CD163, and factor XIIIa.

Aortic involvement is common and consists of circumferential periadventitial infiltration, with possible ostial stenosis in the visceral branches. Cardiac involvement can be found in the coronary arteries, pericardium, or atrial wall. The prognosis is determined by the cardiovascular complications.

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