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On the Cover

What's the diagnosis?

A 73-year-old woman with controlled hypertension and CKD (3B) insidiously developed abdominal and loin discomfort, weight loss, resistant hypertension and rapid decline of GFR. The physical examination was remarkable for hypertension and low-grade fever. A PET-CT scan unmasked a hypermetabolic lesion of the abdominal aorta at the level of D12-L3, further characterized on MRI (Figure 1) as an aortic wall irregularity causing luminal occlusion; the lesion extended to the renal ostia (mainly the left) and the kidneys were asymmetrical and poorly differentiated.

Differential diagnosis was vasculitis, atherosclerosis and rarer aortic neoplasm. The patient was considered unsuitable for endovascular procedure or surgery, given the extension of the disease. An empirical course of corticosteroids for vasculitis was tried. The patient deteriorated with hemodialysis dependent kidney failure, liver and intestinal ischemia and succumbed within days.

The autopsy study (Figure 2) revealed a vegetative and necrotic aortic neoplasm arising from the intima, with involvement by the contiguity of left renal artery; on hematoxylin and eosin stain (Figure 3), the luminal surface had loosely cohesive cells forming aggregates, with a high nuclei/cytoplasm ratio, scarce cytoplasm and marked pleomorphism. There was hepatic, intestinal and pulmonary microembolization.

Malignant renal artery stenosis is rare and has been described in patients with myeloproliferative neoplasms, retroperitoneal sarcoma and aortic intimal sarcoma. Clinical presentation is variable, often indistinguishable from infectious and noninfectious aortitis or atheroembolic disease. The radiomorphological pattern is nonspecific and diagnosis is commonly achieved only after resection. The prognosis is poor. When dealing with abdominal pain, uncontrolled hypertension and kidney failure, vascular disease should be suspected and ruled out.

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(Images and text provided by Telma Santos, Centro Hospitalar e Universitario de Coimbra EPE - Nephrology, Coimbra, Portugal; João Fraga, Centro Hospitalar e Universitario de Coimbra EPE - Pathology, Coimbra, Portugal; Carolina Figueiredo, Centro Hospitalar e Universitario de Coimbra EPE - Nephrology, Coimbra, Portugal; Hélder Moreira, Centro Hospitalar e Universitario de Coimbra EPE - Pathology, Coimbra, Portugal; Andreia Borges, Centro Hospitalar e Universitario de Coimbra - Nephrology, Coimbra, Portugal; Helena Sá, Centro Hospitalar e Universitario de Coimbra - Nephrology, Coimbra, Portugal; and Rui Aloes, Centro Hospitalar e Universitario de Coimbra EPE - Nephrology, Coimbra, Portugal)