ANSWER TO PHOTO QUIZ (PAGE 458)

FATAL SUDDEN PARALYSIS OF THE LOWER EXTREMITIES

DIAGNOSIS

The CT scan revealed a large thrombus in the infra-renal abdominal aorta, the iliac arteries and the upper portion of both femoral arteries (*figure 1A*). Renal and hepatic infarction was seen (*figure 1B*). Combined with the acute presentation and extreme rhabdomyolysis, indicated by the increased CK, ASAT and LDH levels, the diagnosis of acute aortic occlusion (AAO), also known as Leriche syndrome, was made. Aortic occlusion is an extremely rare disease, only a few hundred cases have been reported in the literature. Chronic aortic occlusion usually presents with slowly progressive and vague symptoms such as erectile dysfunction, hypertension, intermittent claudication and symptoms of arterial insufficiency in the lower extremities or even more subtly with chronic deterioration of kidney function alone.[1,2] The typical patients are males between 50-70 years with marked atherosclerosis and a history of smoking. In these patients collateral arteries have been formed, explaining the often mild symptoms. The vascular emergency of AAO, however, typically presents with acute and painful cold ischaemia and rhabdomyolysis.[3,4] A case series from a tertiary hospital, describing 46 consecutive patients with AAO in a period of 40 years, found cardiac embolism to be the cause of AAO in 65%. Atherosclerosis accounted for the remaining 35%. High rates of morbidity and mortality after surgical treatment are described, 74% and 35%, respectively. Recurrent arterial embolism occurred in 43% of the patients with embolic AAO.[5]

In our case, the painless presentation of paralysis and the young age of the patient in the absence of a relevant medical history delayed the correct diagnosis. A broad differential diagnosis, including dry Beriberi, Guillain-Barre, transverse myelitis and polymyositis was made. To exclude these diagnoses, a lumbar puncture was performed, followed by an MRI of the brain and the myelum. It was not until the following morning that a vascular problem was suspected when an ultrasonic arterial Doppler examination showed no arterial pulsations in the femoral arteries, and subsequent CTA of the whole body was carried out, which provided the diagnosis. The patient underwent amputation of both legs combined with aortic thrombectomy to achieve revascularisation: the standard therapy in both acute and chronic Leriche syndrome in the presence of organ or muscular necrosis.[1] His postoperative recovery was complicated by multiple wound infections and decubitus requiring multiple operations and progressive organ failure. The patient opted for euthanasia and received this on the 18th day of admission. Autopsy and hypercoagulability tests, including JAK2 V617F mutation analysis, rotational thromboelastometry, anticardiolipin antibodies, lupus anticoagulants and ANCAs, provided no cause for this fulminant presentation. No structural abnormalities of the heart were found.[6,7]

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