Case Report

The walking man with a completely occluded aorta

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Introduction

Complete aortic occlusion is rare and potentially detrimental. It usually occurs in patients with advanced aorto-iliac atherosclerosis, and can cause severe ischaemic manifestations in the lower extremities, spinal cord, intestine or kidneys [1]. However, the diagnosis may evade detection since collateral vasculature can maintain a basal perfusion and prevent the expression of acute ischaemic phenomena for a long time.

We describe a 69-year-old man with chronic severe aorto-iliac atherosclerotic occlusive disease, who was referred to us late, only after complete suprarenal aortic occlusion had caused acute anuria. This delay limited therapeutic options and aggravated his prognosis.

Case

The patient was referred to our department with anuria that had started abruptly 4 days before admission. Remarkable in his medical history were: cigarette smoking over the last 55 years, acute myocardial infarction 10 years ago, leg claudication over the last 7 years and sudden occurrence of uncontrolled arterial hypertension over the last 2 months. One month before the anuria occurred, the patient was checked by his physician, who diagnosed renal failure (Scr = 1.6 mg/dl, Clcr = 28.9 ml/min/1.73 m²) and arterial hypertension (blood pressure 200/105 mmHg) and prescribed furosemide and diltiazem. No flank pain or haematuria was reported at the time. Until the development of the current anuria, the patient did not have a reduction in urine volume.

On admission, the patient had complete anuria. A harsh systolic murmur was heard over the aortic valve, the carotids and the femoral arteries. A murmur was heard as well as over the abdominal aorta, but only at supraumbilical levels, with abrupt interruption of the murmur below the umbilicus. The pulses were normal over the radial arteries and palpable but less strong over both femoral arteries. The pulses over dorsalis pedis arteries were barely palpable. The systolic blood pressure was measured with a Doppler ultrasound and averaged 170 mmHg over the brachial arteries in both arms and 75 mmHg over the dorsalis pedis arteries in both legs. Nevertheless, no signs of acute lower extremity ischaemia were noted, and despite intermittent claudication, the patient could still walk. The rest of the physical examination was normal. The laboratory test results were as follows: Ht = 28.7%, Hb = 9.5 g/dl, WBC = 5400/μl, PLT = 58 000/μl, PT = 18 s, PTT = 27 s, D-dimers = 1000, fibrinogen = 221 mg/dl, serum urea = 141 mg/dl, creatinine = 7.5 mg/dl, ESR = 65 mm/h, CRP = 33.5 mg/l. Tests for a hypercoagulable state were negative as were the cancer indices. Intensive work-up failed to reveal any underlying tumour or systemic disease. Ultrasonography revealed a smaller right kidney (9.4 cm, cortex ~8 mm) than the left one (10 cm, cortex ~10 mm), with no signs of obstruction in either kidney. The diagnosis of acute anuric renal failure, presumed to be because of chronic ischaemic nephropathy, was made, and the patient started haemodialysis. A ⁹⁹ᵐ-Tc DTPA nephrogram vaguely visualized both kidneys, which appeared minimally functional. On ⁹⁹ᵐ-Tc MAG3 scintigraphy, the curves of both kidneys appeared flattened, while scintigraphic aortography showed total interruption of the perfusion of the aorta, at a suprarenal level, and of both renal arteries. A CT revealed the absence of radiocontrast media in the aorta from below the Haller tripod down to the bifurcation of the iliac arteries, due to thrombosis, and no retroperitoneal adenopathy. Aortography showed complete occlusion of the aorta from below the celiac artery.
down to the femoral arteries, with extensive collateral circulation through the superficial circumflex iliac arteries and the superficial epigastric arteries, and also the occlusion of the superior mesenteric artery, the distal branches of which were visualized through the pancreatoduodenal circulation (Figure 1). A Triplex study of the legs revealed perfusion of the iliac, femoral and popliteal arteries and veins (Figure 2). The treatment proposed to the patient was surgical thrombectomy in the abdominal aorta and endarterectomy of renal arteries—a recommendation that the patient, surprisingly, rejected. He continued on haemodialysis for 3 months, when he died of ischaemic intestinal necrosis and sepsis.

Discussion

Complete aortic occlusion is rare but potentially catastrophic. Acute aortic occlusion bears an early mortality of 31–52% [1] and is caused either by embolic occlusion of the infrarenal aorta at its bifurcation, known as a ‘saddle embolus’, or by acute thrombosis of the abdominal aorta. Between 75 and 80% of cases of thrombotic aortic occlusion occur in the setting of underlying severe aorto-iliac atherosclerotic occlusive disease, often precipitated by a low-flow state secondary to heart failure or dehydration. In the rest, a hypercoagulable state may precipitate thrombosis of an abdominal aortic aneurysm and lead to aortic occlusion [2]. In a study by Tapper et al. [3], 12% of patients with aortic occlusion had suprarenal aortic occlusion; 81.2% of this group had chronic aortic occlusion with proximal propagation of thrombus to involve the suprarenal aorta. The main manifestations in these patients were uncontrolled hypertension and claudication. An early diagnosis of aorto-iliac disease is necessary in order to avoid the fatal outcome. A simple, readily available and non-invasive approach to diagnosis is duplex scanning and sonography of the aorta, iliac and common femoral arteries. In severe occlusive disease, its sensitivity reaches 91% and its specificity 93% [4]. It has been shown that duplex scanning can efficiently detect the occlusion and visualize collateral circulation [5,6], and is often used as the sole preoperative method of visualizing the aorto-iliac segment [4,7], in some cases making angiography redundant. For suprarenal aortic occlusion two therapeutic options exist: for patients with a short life expectancy with a rather stable renal function (through collateral perfusion) and controllable hypertension, palliative axillo-bifemoral bypass is the treatment of choice [8]. Most patients, however, will require aortic reconstruction and thrombectomy of the juxtarenal segment [8,9]. Cases of acute renal failure caused by aortic thrombosis have been treated with success with the reversion of the thrombus, even after several days of anuria [10].

In our patient, the history (heavy smoker over the last 55 years, intermittent claudication over the past 7 years), clinical findings (murmurs over all major arteries, with palpable pulses over both the femoral arteries and absence of signs of acute ischaemia) and the extended collateral circulation, suggest a chronic, severe aorto-iliac atherosclerotic occlusive disease while the smaller size of the right kidney speaks for ischaemic nephropathy. Moreover, the patient’s haematological profile (thrombocytopenia, increased PT and D-dimers and decreased PTT and fibrinogen) favours the diagnosis of chronic disseminated intravascular coagulopathy, probably due to the chronic presence of a large thrombus in the aorta. The sudden manifestation and diagnosis of severe arterial hypertension and renal dysfunction 1 month before admission, suggest a proximal propagation of the aortic thrombus. Early duplex scanning and sonography of the aorta at that time would have been adequate to timely detect collateral circulation and aortic occlusion. This delay
in the diagnosis was critical in our patient and aggravated his prognosis, for abrupt anuria supervened a few days before admission, at a time when the thrombus had also involved the suprarenal aorta up to the superior mesenteric artery. A serious drawback of the diagnostic work-up was the use of iodinated contrast media to diagnose the nature and visualize the exact extent of the aortic occlusion, so as to select the appropriate treatment. Magnetic resonance angiography in these cases is a much safer procedure for the preservation of renal function. In urgent cases when MRI angiography is unavailable, as in our case, the use of one of the new generation of iodinated contrast materials for angiography is unavoidable.

In cases of severe chronic aorto-iliac atherosclerotic occlusive disease, an extended collateral circulation develops [11]. So too in our patient; despite severe chronic occlusive disease, an adequate basal perfusion of the intestine and lower extremities was maintained for a long time, due to the extended collateral vascular network, which bypassed the superior mesenteric artery, the occluded part of the aorta and the iliac arteries. This circulation protected the patient from acute ischaemic manifestations for a long time, and that probably was the main reason for the late referral of the patient to a central medical facility. Had the patient been worked-up at the time of the first diagnosis of hypertension or renal failure, before he manifested acute anuria, he could have been a candidate for a timely and relatively safer endovascular approach or thrombectomy, given the fact that the operative mortality rate in chronic infrarenal aortic occlusion is \( \approx 5\% \) [12] while in chronic suprarenal aortic occlusion it is 23% [3]. Be that as it may, the patient rejected any surgery, and had a course that was predetermined, given the severity of his condition.

In conclusion, aorto-iliac occlusive disease causing ischaemic nephropathy should be suspected in patients who suddenly manifest uncontrollable arterial hypertension or renal function deterioration, especially after the use of ACE inhibitors or diuretics, when other predisposing factors coexist (smoking, male sex, hyperlipidaemia, chronic renal failure) or when atherosclerotic disease (intermittent claudication, coronary artery disease, cerebrovascular disease) is also evident. A thorough clinical and laboratory evaluation of such patients should be performed, since symptoms alone can be misleading, due to the development of extended collateral vasculature, which can prevent the manifestation of acute ischaemic phenomena. Duplex scanning and sonography of the aorta and iliac arteries are simple examinations that can certainly help make an early diagnosis. Immediate angioplasty or thrombectomy is obligatory when aortic occlusion is diagnosed. Without surgical treatment, patients with aortic occlusion have a poor prognosis.

References


Conflict of interest statement. None declared.