Case Report

Cholesterol embolism associated with macroscopic renal infarction

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Introduction

Atheromatous renal disease is the major cause of renal insufficiency in the elderly, and cholesterol embolism is a manifestation of this disease. Cholesterol embolism occurs in patients suffering from diffuse erosive atherosclerosis, usually after triggering causes, such as aortic surgery, arterial invasive procedures (angiography, left heart catheterization and coronary angioplasty) and anticoagulant therapy [1]. In addition, it can also be observed in patients with acute myocardial infarction to whom thrombolytic agents have been administered [2]. Moreover, a case of cholesterol emboli syndrome combined with acute interstitial nephritis after thrombolytic therapy with streptokinase recently has been described [3]. On the other hand, cholesterol embolism may also occur spontaneously. The incidence of the spontaneous form is difficult to determine, but in a study performed by Cross et al. in the UK, 2% of 372 necropsies showed cholesterol emboli in spleen or kidneys [4]. In this study, age and sex distribution were similar to that reported in the population with atherosclerosis, and middle-aged men were predominantly affected. The spectrum of 87 diseases caused by cholesterol emboli ranges from asymptomatic to rapidly progressive multiple system failure which is associated with a mortality rate of > 80% [5]. The initial signs and symptoms in a study of 33 patients diagnosed with cholesterol embolism were blue toes syndrome, livedo reticularis, gangrene, leg, toe or foot pain, abdominal pain and flank or back pain, gross haematuria, accelerated hypertension and renal failure. Unusual presentations were spinal cord infarction, non-healing foot ulcers, penile gangrene and haematochezia [6]. Also, endogenous lipid pneumonia has been described as a specific pulmonary involvement [7]. Cholesterol embolism may also be associated with fever, increased erythrocyte sedimentation rate and eosinophilia. Thus, in cases of spontaneous cholesterol embolism, differential diagnosis includes, polyarteritis nodosa, allergic vasculitis and subacute bacterial endocarditis [8].

We describe a case of spontaneous cholesterol emboli syndrome associated with macroscopic renal infarction, severe systemic arterial hypertension and rhabdomyolysis.

Case

A 62-year-old male was admitted to another hospital because of sudden and piercing lumbar pain and paraplegia. He had given up smoking 2 years previously. The patient had a 1 year history of intermittent claudication, but he did not ask for a medical evaluation for it. The current clinical picture started when he experienced pain in both calves while he was walking swiftly. The pain did not stop when he rested. The pain grew in intensity and turned into severe and excruciating lumbar pain with belt-type irradiation. He was unable to walk anymore and progressive numbness developed in both legs. Physical examination revealed arterial hypertension (210/130 mmHg), paraparesis 1-2/5, tendon arreflexia, absent position and vibratory sensation in the lower limbs with greatly increased muscle tone. Peripheral pulses were present. Skin lesions in the form of livedo reticularis appeared on both legs, progressing to the trunk. An abdominal ultrasound disclosed an aneurysmatic aorta. With the initial diagnosis of dissecting aortic aneurysm, sodium nitroprusside was initiated and the patient was transferred to our hospital. On arrival, the physical examination displayed arterial hypertension (190/100 mmHg), sinus tachycardia, normal cardiac and pulmonary sounds, and livedo reticularis involving both legs and inferior abdomen. The patient was alert and orientated, complaining of excruciating lumbar pain with posterior irradiation to both legs. Cranial nerve functions were preserved. There was a symmetrical paraparesis 1-2/5. There was not a sensory level. Optic fundi revealed diminished retinal arteriole calibre with a microinfarction close to the left papilla and
sclero-hypertensive changes in both eyes: intra-arteriole cholesterol clefts were absent. Laboratory studies showed: serum creatinine 199 μmol/l, serum urea 37.3 mmol/l, total serum creatine-kinase 800 U/l (normal range 12–70 U/l), alanine aminotransferase 10.8 μkat/l (normal range 0.5–0.77 μkat/l). In serum, aspartate aminotransferase, ɣ-glutamyltransferase, alkaline phosphatase, bilirubin, amylase, calcium, sodium and potassium were normal. Human immunodeficiency virus, syphilis and hepatitis (B and C) serologies, as well as rheumatoid factor, complement fractions, cryoglobulins, antinuclear antibodies and antineutrophil cytoplasmic antibodies were negative. There was no anaemia, and the white cell count was 17 500/mm³ with neutrophilia but not eosinophilia. The erythrocyte sedimentation rate was 28 mm/h. The platelet count was 175 000/mm³ with normal fibrinogen, protrombin and tromboplastin times. Blood cultures were negative. Urinalysis revealed a positive myoglobin urinary test, moderate proteinuria and microhaematuria. Electrocardiogram and chest radiographs were normal. A thorax scan showed a non-fissured aneurysm of the ascendent aorta and a non-fissured abdominal aortic aneurysm with a parietal thrombus running down from the ostium of the renal arteries, as well as a right kidney hypoperfusion image suggesting renal infarction (Figure 1). No images of aortic dissection were observed.

The patient’s neurological condition worsened rapidly, dysarthria appeared, the patient became increasingly agitated and, finally, mechanical ventilation was required. The neurological findings were consistent with ischaemic disturbance involving mainly the lumbo-sacral anterior and posterior spinal cord, along with the central nervous system. Vascular surgery of aortic aneurysms was not indicated. Arterial hypertension was controlled with nifedipine. The oliguric acute renal failure initially was attributed to rhabdomyolysis and managed by means of haemodialysis. Bilateral amputation of the lower limbs was required because of the progression of the ischaemic lesions to gangrene. The patient’s status worsened and he died on the 15th day after admission due to a septic process secondary to aspiration pneumonia. A post-mortem renal biopsy performed on the left kidney was diagnostic of cholesterol microembolism (Figure 2).

Discussion

We have described a case of a fatal spontaneous cholesterol embolism affecting the kidneys, as demonstrated by renal biopsy, and probably the spinal cord, central nervous system and muscular peripheral vessels. To our knowledge, the association of cholesterol embolism with macroscopic renal infarction has not been reported previously. This fact made us review the spontaneous origin of this event, even though it occurred in the absence of any of the described triggering factors for cholesterol embolism.

Embolism of cholesterol crystals may be a casual autopsy finding, may produce blue toe syndrome or livedo reticularis, or may evolve, as in our patient, in the form of multiorgan life-threatening event. Multisystemic signs and symptoms led us to consider a diagnosis different from systemic vasculitis. In some instance, this differential diagnosis may be difficult, since cholesterol microemboli syndrome can be associ-
ated with necrotizing immunonegative glomerulonephritis [9]. In our case, the negative antineutrophil-cytoplasmic antibodies (ANCA) and particularly the absence of glomerular or vasculitic lesions in the renal biopsy ruled out the diagnosis of systemic vasculitis.

The absence of a precipitating event made the diagnosis of cholesterol emboli syndrome more difficult. In some cases, the previous medical history of vascular disease (as in our patient), the demonstration of ulcerative atheromatous lesions of the thoracic aorta by transoesophageal echocardiography (10) and eosinophilia, eosinophiluria and hypocomplementemia (absent in our case) may help in the diagnosis [11]. In our case, there was a macroscopic renal infarction, probably caused by dislodgement of an aortic atheromatous plaque or by a clot from the thrombosed aortic aneurysm. The renal infarction may explain, at least in part, the sudden lumbar pain experienced by the patient, the microhaematuria and the arterial hypertension, that, in this context, is usually secondary to an increased renin release [12]. The extensive array of neurological signs and symptoms described may reflect the concomitant macroembolization and microembolization of the cerebral and spinal vasculature. On the other hand, rhabdomyolysis was also probably derived from muscle ischaemia. The presence of peripheral pulses suggests that muscle microvasculature embolization by aortic debris was mainly involved in this event.

Three different and severe renal insults were detected in this case: (i) macroscopic infarction of the right kidney; (ii) renal tubular toxicity caused by rhabdomyolysis; and (iii) cholesterol crystal embolization as revealed by post-mortem renal biopsy. As a possible pathogenic mechanism, we propose that the renal infarction caused an acute renovascular arterial hypertension episode that resulted in a massive release of cholesterol crystals from the plaques of the atheroma, thus affecting the kidneys, skin, muscles and nervous system. Therefore, in cases of spontaneous cholesterol embolism, it would be relevant to look for the presence of concomitant renal macroscopic infarction.

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