Case Report

Is atheroembolic disease a new differential diagnosis of pulmonary–renal syndrome?

Didier Ducloux, Valérie Schuller, Elisabeth Ranfaing, Véronique Fournier, Jean-Michel Rebibou, Laurent Martin and Jean-Marc Chalopin

1Department of Nephrology and Renal Transplantation, Hopital Saint Jacques, 2Department of Pathology, Hopital Minjoz; Besançon, France

Introduction

Atheroembolic disease is an infrequent disorder that occurs as a result of embolization of cholesterol crystals from atheromatous plaques lining the aorta and other major arteries. The clinical presentation is often characteristic and may include livedo reticularis, gangrenous extremities, renal failure, hypertension, and pancreatitis. Intestinal involvement with abdominal, nausea, and gastrointestinal bleeding is occasionally the clinical presentation, but frank infarction of the small and large bowel is rare [1,2].

Pulmonary involvement observed during atheroembolic disease is usually due to congestive heart failure and not to pulmonary cholesterol crystal emboli. Very few cases of biopsy-proven specific pulmonary involvement have previously been reported in the course of atheroembolic disease [3,4]. We describe a case of atheroembolic disease associated with endogenous lipoid pneumonia. Because spumous macrophages are observed in the inflammatory reaction surrounding cholesterol crystal clefts in different sites, we hypothesize that their presence in brochoalveolar fluid may reflect the presence of cholesterol crystal emboli in the pulmonary parenchyma.

Case report

A 72-year-old man was admitted to the hospital because of acute renal failure. The patient had a 20-year history of hypertension and had peripheral vascular disease. During the year before admission, the serum creatinine concentration had ranged between 112 and 156 μmol/l. Anamnesis revealed that he had had an aortography by femoral arterial puncture 2 months before. There was no history of recent fever, haematuria, or diabetes mellitus, and no family history of renal disease. The temperature was 37°C, the pulse was 90/min and the blood pressure 170/100 mmHg. Physical examination revealed purple toes and livedo reticularis. Significant laboratory data were as follows: serum creatinine concentration, 356 μmol/l; white blood cell count, 12 000/mm³ with eosinophil polymorphonuclear, 1670/mm³; haemoglobin concentration, 136 g/l. Radiography of the chest was normal. Atheroembolic renal disease was suspected and a skin biopsy was performed. Histological examination revealed intra-arteriolar cholesterol crystal clefts (Figure 1). An osteomediadary biopsy revealed eosinophilia and the presence of lipophages. Renal failure worsened and a peritoneal catheter was inserted.

Ten days after admission the patient became dyspnoeic. He was able to lie flat and had no peripheral oedema, fever, sweats, chills, or cough. Inspiratory crackles were heard over the lower one-third of both lungs. Echocardiography was normal. Arterial-blood gases were the following while breathing oxygen at 6 litres per minute: partial pressure of oxygen, 8 kPa; partial pressure of dioxide 4.5 kPa; pH 7.35. A radiograph of the chest showed bilateral infiltrates more prominent in right mid- and lower lung fields. Computed tomography revealed extensive bilateral infiltrates. Flexible fibroptic bronchoscopy examination did not reveal endobronchial lesion. A bronchoalveolar lavage was performed. Bronchoalveolar fluid was sterile and microscopic examination revealed no acid-fast bacilli. Bronchoalveolar fluid analysis was positive for lipophages (Figure 2). The patient did not receive either vegetable or mineral oils.

The patient improved slightly without any specific treatment. No diuretic therapy was instituted, and weight remained stable during clinical and radiological improvement. Peritoneal dialysis was begun 2 weeks later.

Discussion

Our observation describes the occurrence of a lipoid pneumonia in a man with atheroembolic disease. Atheroembolic disease was suspected because of recent...
arterial manipulation, clinical presentation and hyper-
3 eosinophilia, and confirmed by skin biopsy exhibiting
cholesterol crystal clefts.

Pulmonary oedema is the main cause of pulmonary
involvement in atheroembolic disease. Indeed hyper-
tension, volume overload, and cardiac failure are fre-
quent in atheroembolic disease and may contribute to
pulmonary oedema. Although we did not perform
right cardiac catheterization, pulmonary oedema is
unlikely in our patient. He was not orthopnoeic and
had neither oedema nor symptoms of heart failure. No
diuretic therapy was instituted and weight remained
stable during clinical and radiological improvement.
Moreover, echocardiography was normal.

Patients hospitalized because of acute renal failure are
prone to infectious complications. Nevertheless, because
both bronchoalveolar fluid and blood cultures remained
sterile, an infectious pneumonia is also unlikely.

There is some evidence that our patient exhibited
lipid pneumonia. Indeed, clinical presentation and
radiological findings are concordant with previous
descriptions [5]. Moreover, the presence of numerous
lipophages (>5%) in bronchoalveolar fluid confirmed
the diagnosis [6].

Exogenous lipid pneumonia, which results from
aspiration of oil, can be ruled out since our patient
did not receive either vegetable or mineral oils [6]. A
self-medication is excluded because of the patient’s
disability. Moreover, the presence of lipophages in
bone marrow suggests an endogenous mechanism for
the presence of lipophages in lungs. Endogenous lipid
pneumonia includes cholesterol pneumonitis, fat
embolism, pulmonary alveolar proteinosis, and lipid
storage disease [7]. Alveolar proteinosis is unlikely in
our patient. Indeed alveolar proteinosis is a progressive
disease, and spontaneous clinical and radiological
remission is not a feature [8]. Moreover bronchoalveo-
lar fluid analysis was not consistent with alveolar
proteinosis. Other differential diagnoses are very rare
and characteristic enough to be excluded in this case.

Our patient had both atheroembolic disease and
endogenous lipid pneumonia of unknown origin. We
suspect lipid pneumonia to be due to cholesterol
emboli in the lungs. Firstly cholesterol crystals may
embolize from aorta through the bronchial arteries in
the pulmonary parenchyma. Secondly spumous macro-
phages are observed in the inflammatory reaction
surrounding cholesterol crystal clefts. Lipophages
Fig. 1. Cutaneous biopsy (H&E Safran) exhibiting inflammatory
reaction surrounding intra-arteriolar cholesterol emboli.

Fig. 2. Bronchoalveolar fluid (Soudan Rot B) exhibiting numerous
lipophages.
Is atheroembolic disease a new differential diagnosis of pulmonary–renal syndrome?

observed in bronchoalveolar fluid are likely to reflect the local inflammatory response to cholesterol emboli. Finally it is difficult to accept that the endogenous lipoid pneumonia represented a chance occurrence in a patient already affected by a rare disease.

To our knowledge, only two cases of pulmonary atheroembolic disease have been previously reported [3]. Scully et al. described a man with atheroembolic disease who developed acute pneumopathy. Open-lung biopsy showed atheromatous emboli in a quarter of the pulmonary arteries. The emboli had elicited an inflammatory reaction in the adventitia of muscular pulmonary arteries and cholesterol crystals were surrounded by histiocytes. Eighteen years earlier, a similar case had been published [4]. The authors reported the case of a man who died from respiratory failure 15 days after aortic surgery. Autopsy showed that the cause of death was widespread atherosclerotic emboli from the aorta to the pulmonary circulation.

In conclusion we think that cholesterol emboli may induce specific pulmonary involvement. Bronchoalveolar fluid analysis demonstrating lipophages may allow rapid and easy diagnosis. Atheroembolic disease should be considered as a new cause of pulmonary–renal syndrome.

References


Received for publication: 25.4.97
Accepted in revised form: 15.10.97