Renal-limited polyarteritis nodosa presenting with loin pain and haematuria

J. L. Górriz¹, A. Sancho¹, R. Ferrer², E. Alcoy¹, J. F. Crespo¹, J. Palmero³ and L. M. Pallardó¹

Departments of ¹Nephrology, ²Pathology, and ³Radiology, Hospital Universitario Dr. Peset, Valencia, Spain

Key words: haematuria; hypertension; kidney failure; loin pain; polyarteritis nodosa; vasculitis

Introduction

Classic polyarteritis nodosa (PAN) is a medium-sized vessel vasculitis with multiorgan involvement [1]. Its clinical polymorphism renders diagnosis difficult, especially in cases with uncommon presentation [1–3]. We report a case of polyarteritis nodosa with renal-limited involvement presenting with haematuria, loin pain, and subacute renal failure.

Case report

A 49-year-old Caucasian man with a previous history of long term headache and left renal lithiasis treated with extracorporeal lithotripsy was seen at emergency room because of severe loin pain as well as macroscopic haematuria. Hypogastric and left lower quadrant pain was also noted. Blood pressure was 180/110 mmHg, and abdominal tenderness was detected. Apart from macroscopic haematuria there were no relevant laboratory findings, with a plasma creatinine level of 97 μmol/l. A diagnosis of renal colic was made, which improved with analgesic treatment. He was also given nifedipine as an antihypertensive treatment. One month later he returned to hospital because abdominal loin pain and macroscopic haematuria reappeared, showing renal functional impairment (urea nitrogen 12 mmol/l, creatinine 185 μmol/l). He looked healthy, his temperature was 36.4°C and his blood pressure was 180/110. On physical examination, abdominal tenderness in the hypogastrium was detected. No abdominal bruits were heard. No other abnormal physical findings were noted. Progressive renal function impairment was observed in the following 2 days (urea nitrogen 23 mmol/l, creatinine 354 μmol/l). In order to control blood pressure atenolol 25 mg daily, nifedipine 30 mg twice a day, and enalapril 20 mg daily were given. Other laboratory studies showed phosphorus 5.7 mg/dl and haemoglobin of 14.0 g/dl. The remaining parameters were normal, including bilirubin, glucose, serum proteins and electrophoresis, aminotransferases, cholesterol, triglycerides, creatine kinase, amylase, carbon dioxide, coagulation tests, white-cell and differential count. Serological evaluation for immunoglobulins, complement, ANA, anti-DNA, anti-globular basement membrane antibodies, ANCA, anti-ENA, cryoglobulins, circulating immune complexes, anticoagulins, hepatitis B and C viruses and HIV antibodies were either normal or negative. Plasma renin activity was 16 ng/ml/h (normal 0.5–2.8), plasma aldosterone 477 mg/ml (normal < 150). Proteinuria was 0.6 g/day. Urinary analysis showed nephritic sediment, with numerous erythrocyte and granular casts, and low-grade microhaematuria/leukocyturia. Urine culture was negative. Abdomen and chest X-ray films, fundoscopy and electrocardiogram were normal. Abdominal and renal ultrasonographic study, including renal artery Doppler sonogram, showed no abnormalities.

Because of the presence of subacute renal failure, a percutaneous renal biopsy was performed, producing 11 glomeruli that were histologically normal. Interstitium and tubules were unremarkable except at one site, where interstitial oedema and an artery with moderate neutrophilic and eosinophilic infiltration of the adventitial and medial layers were seen, besides reduplicated and ruptured elastic fibres. Focal fibrinoid intramural deposition was found. Immunofluorescence microscopy was negative. An angiographic study showed multiple microaneurysms of the renal vasculature in both kidneys (Figure 1) and a large aneurysm in the left renal artery. Supra-aortic branches and mesenteric arteries showed no aneurysms.

A diagnosis of PAN was made and three intravenous methylprednisolone pulses (1 g × 3 days) were given, followed by prednisone at initial dose of 1 mg/kg/day. Six monthly cyclophosphamide courses (0.5 g/m²) were also administered. A routine renal Doppler ultrasonography 1 week after biopsy revealed an arterio-
usually related to increased levels of plasma renin activity [7]. The diagnosis can be established by histology, e.g. biopsy of an affected organ, usually striated muscle [8], or by renal biopsy. The hallmark is involvement of medium-sized arteries. Another typical sign is the presence of aneurysms on arteriography [1,8].

Our case was remarkable because he presented with renal manifestations only, i.e. haematuria, loin pain, hypertension and subacute renal failure. The misdiagnosis of renal colic was made because of flank pain and a history of renal lithiasis. Progressive renal function impairment at the time of the second admission led to reversal of the diagnosis. Renal biopsy strongly suggested the possibility of PAN, a diagnosis which was confirmed by arteriography.

In our patient high renin plasma levels were detected, as has been described by other authors, related to focal ischaemia, which causes activation of the renin–angiotensin system analogous to the changes caused by renal artery stenosis [7]. Thus subacute renal failure with high plasma renin in the absence of malignant hypertension or bilateral renal artery stenosis is consistent with the possibility of PAN.

Although ANCA were negative in our patient, they are present in 25–50% of PAN patients [9]. The prevalence is lowest in cases that are not associated with hepatitis B virus infection [4]. Nevertheless, the use of differing diagnostic criteria in PAN studies reported in the literature makes it difficult to assess the true prevalence of ANCA in these patients.

PAN treatment includes corticosteroids, but most of authors advocate adding a cytotoxic agent, usually cyclophosphamide [10]. The addition of plasmapheresis to corticosteroids and cyclophosphamide was not followed by significant differences in the 5-year patient survival [8]. The outcome was poor in untreated patients and in those with impaired renal function on admission [8]. So early diagnosis and treatment are crucial. In fact the delay of a month to establish the correct diagnosis in our patient led to severe renal functional impairment, which fortunately was reversible. Although an arteriovenous fistula can be a complication of a renal biopsy, an increased risk of bleeding and arteriovenous fistulae formation has been reported in PAN patients [11]. In our case an arteriovenous fistulae was detected after the kidney biopsy, but it healed spontaneously.

In conclusion, PAN may have an uncommon presentation as a renal-limited form presenting with loin pain and haematuria.

Discussion

PAN is a necrotizing vasculitis of medium-sized vessels. Its origin is unknown. It may have an acute or subacute course [4] and cause formation of multiple aneurysms [1]. The clinical picture is characterized by fever and malaise, with peripheral neuropathy, arthralgia, myalgia, and visceral involvement (gastrointestinal tract, kidneys, and heart) [1]. Renal involvement occurs in 80–90% of patients, characterized by haematuria and loin pain due to renal infarction or renal impairment related to ischaemia [5]. It is usually a systemic disease and the renal-limited form is rare [6]. Hypertension is common, affecting 25–71% of patients [1], and it is usually related to increased levels of plasma renin activity [7]. The diagnosis can be established by histology, e.g. biopsy of an affected organ, usually striated muscle [8], or by renal biopsy. The hallmark is involvement of medium-sized arteries. Another typical sign is the presence of aneurysms on arteriography [1,8].

Our case was remarkable because he presented with renal manifestations only, i.e. haematuria, loin pain, hypertension and subacute renal failure. The misdiagnosis of renal colic was made because of flank pain and a history of renal lithiasis. Progressive renal function impairment at the time of the second admission led to reversal of the diagnosis. Renal biopsy strongly suggested the possibility of PAN, a diagnosis which was confirmed by arteriography.

In our patient high renin plasma levels were detected, as has been described by other authors, related to focal ischaemia, which causes activation of the renin–angiotensin system analogous to the changes caused by renal artery stenosis [7]. Thus subacute renal failure with high plasma renin in the absence of malignant hypertension or bilateral renal artery stenosis is consistent with the possibility of PAN.

Although ANCA were negative in our patient, they are present in 25–50% of PAN patients [9]. The prevalence is lowest in cases that are not associated with hepatitis B virus infection [4]. Nevertheless, the use of differing diagnostic criteria in PAN studies reported in the literature makes it difficult to assess the true prevalence of ANCA in these patients.

PAN treatment includes corticosteroids, but most of authors advocate adding a cytotoxic agent, usually cyclophosphamide [10]. The addition of plasmapheresis to corticosteroids and cyclophosphamide was not followed by significant differences in the 5-year patient survival [8]. The outcome was poor in untreated patients and in those with impaired renal function on admission [8]. So early diagnosis and treatment are crucial. In fact the delay of a month to establish the correct diagnosis in our patient led to severe renal functional impairment, which fortunately was reversible. Although an arteriovenous fistula can be a complication of a renal biopsy, an increased risk of bleeding and arteriovenous fistulae formation has been reported in PAN patients [11]. In our case an arteriovenous fistulae was detected after the kidney biopsy, but it healed spontaneously.

In conclusion, PAN may have an uncommon presentation as a renal-limited form presenting with loin pain and haematuria.

References

3. Perirenal haematoma as the presenting feature of polyarteritis
Renal-limited polyarteritis nodosa with loin pain and haematuria


Received for publication: 13.5.97
Accepted in revised form: 11.7.97