Interesting Case

Renal artery dissection secondary to medial hyperplasia presenting as loin pain haematuria syndrome

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Introduction

Fibromuscular dysplasia (FMD) accounts for 5–10% of renal artery stenosis. The majority of cases (90%) involve the media. Loin pain haematuria syndrome is characterized by loin pain, which is often severe and unrelenting, in association with haematuria of glomerular origin. We report a case of medial hyperplasia with dissection of the renal artery, where the patient presented with loin pain haematuria syndrome without hypertension.

Case

A 42-year-old heavy goods vehicle driver presented with bilateral, non-radiating loin pain associated with microscopic haematuria. He underwent normal urological investigation including intravenous urography and cystourethroscopy, renal function was normal as judged by a serum creatinine of 98 mmol/l and he was normotensive. Despite negative investigations he remained symptomatic but was not readmitted until 10 years later when he had a further episode of severe loin pain. At the time of admission he was febrile (38.6°C) but remained normotensive. His lower abdomen was tender and distended. Laboratory investigations revealed a haemoglobin of 14.2 g/dl, white cell count 17.8 × 10⁹/l, serum sodium 139 mmol/l, potassium 4.6 mmol/l, creatinine 103 mmol/l, albumin 4.3 g/dl, protein 7.8 g/dl, bilirubin 11 mmol/l, alkaline phosphatase 109 IU/l, amylase 31 IU/l, aspartate aminotransferase 27 IU/l and C-reactive protein 139 mg/l. Barium enema examination showed no abnormality. He subsequently underwent laparotomy for continuing symptoms, but no pathology was found. Despite negative cultures his symptoms and signs subsequently settled after several days of antibiotic treatment. Following discharge from hospital he continued to experience intermittent bilateral loin pain and 22 months later was admitted to hospital in Germany. Intravenous pyelography revealed minor filling defects within the pelvicalyceal system in the left upper pole, but no significant calyceal distortion or displacement. CT scanning revealed a 4.5 cm mixed density lesion in the upper pole of the left kidney (Figure 1). The right kidney was normal. The patient returned to England and was readmitted 2 months later with severe left loin pain. A radical left nephrectomy was performed for presumed malignancy. The kidney was slightly enlarged (12 × 6 × 5.5 cm, 208 g) and showed patchy, demarcated, irregular yellow areas

Fig. 1. CT scanning revealing a 4.5 cm mixed density lesion in the upper pole of the left kidney.
with associated cortical scarring in the lower pole, suggestive of multiple infarcts of varying age. The rest of the kidney appeared normal. Histology revealed coagulative necrosis of the glomeruli and tubules with a mixed acute and chronic inflammatory response. Immunohistochemistry was negative. The main renal artery showed considerable compression. There was evidence of dissection of the outer media, which showed fibroplastic and smooth muscle proliferation. The adventitia was normal. The appearances were consistent with FMD of the renal artery of medial type, subtype medial dissection. The patient was referred to the renal unit for subsequent follow up. Three months following his left nephrectomy he experienced an episode of right loin pain and dark coloured urine. Renal angiography revealed minimal narrowing of the right renal artery at the point of division of the arteries in the renal hilum. The appearances were consistent with medial hyperplasia (Figure 2). Coeliac axis angiography performed at the same time was entirely normal. He has subsequently remained well with no further symptoms and continues to be normotensive (blood pressure 110/60 mmHg) on no treatment. His chromium-labelled EDTA GFR 14 months post-nephrectomy was 80 ml/min and his serum creatinine level 24 months post-nephrectomy remains normal at 89 mmol/l.

Discussion

Loin pain haematuria syndrome has been a described clinical entity since 1967 and presents with flank pain, haematuria and occasional low-grade pyrexia [1]. Over the years there has been little progress in establishing the aetiology and underlying pathology. Although commoner in females, the syndrome is well described in males; the age at onset varies between early adulthood and middle age. Loin pain haematuria syndrome can cause debilitating symptoms but is not normally associated with development, or progression, of renal functional impairment.

FMD is the second most common cause of renal artery stenosis accounting for <10% of cases. Ninety percent of cases of fibromuscular dysplasia involve the media. It is more common in women than men and in white populations as compared to African-American and Asians. Luscher et al. [2] studied 92 patients with FMD. Half of the patients with bilateral renovascular disease showed additional extrarenal FMD; all patients with renovascular FMD were hypertensive (mean blood pressure 194 ± 34/119 ± 18 mmHg). FMD has been classified based on the layer of the vessel wall affected and is classified in Table 1 [3].

Medial hyperplasia alone accounts for 5–15% of cases of renal FMD and usually occurs in the second decade in women and in the third decade in men. Angiographically, the stenosis is smooth and linear. Many reports have grouped intimal fibroplasia and medial hyperplasia together [4,5].

Medial dissection accounts for 5–10% of cases involving the media. It begins as a defect in the internal

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<td>intimal fibroplasia</td>
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<td>(ii) Media</td>
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<td>(a) medial hyperplasia</td>
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<td>(b) medial fibroplasia with aneurysms</td>
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<td>(c) perimedial fibroplasia</td>
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<td>(d) medial dissection</td>
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<td>(iii) Adventitia</td>
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elastic lamina, allowing the blood to gain access to the media. The channel is usually located in the outer third of media and is surrounded by fibrous tissue and it may be accompanied by intimal fibroplasia [3]. Renal infarcts are more common in this lesion than in the other types of renal artery dysplasia.

Progression of renal artery stenosis varies with the subtype. In one study it was found that medial fibroplasia was least progressive as it burnt out at some stage, while intimal fibroplasia progressed to narrowing to a greater extent [5]. In the second study from the same institution, progression of narrowing occurred in 33% over 4 years, considerably higher than in the previous study [6]. Goncharenko et al. [4] described progression in all subtypes with renal infarction occurring in 24% of patients.

Angiographic abnormalities have been noted on the non-affected side as well as the symptomatic side and follow up renal angiography has suggested progression from local ischaemia to renal scarring [1]. Histopathological examination of nephrectomy specimens has revealed cortical ischaemia, infarction, and intimal lesions from segmental arteries.

Our patient presented with the features of loin pain haematuria syndrome and initial urological investigation was negative, his renal function was normal and he was normotensive. His symptoms subsequently persisted for 12 years prior to nephrectomy. He had remained normotensive at each subsequent hospital visit both prior to and following nephrectomy. The histopathological findings were consistent with multiple infarcts of varying age, together with medial hyperplasia subtype medial dissection. The aetiology of loin pain haematuria syndrome is poorly understood and the diagnosis is made by exclusion of other pathologies resulting in haematuria associated with renal pain. This case illustrates the importance of complete investigation of loin pain associated with haematuria. CT with intravenous contrast has been useful in the diagnosis of renal infarct especially in acute stages of the infarct. As the infarct evolves into tumefactive necrosis the diagnosis becomes more difficult [7,8]. We have reported use of nuclear scanning in a case of segmental renal infarction, where the index of suspicion was high. Nuclear scanning has been described in few case reports to detect infarcted areas [9,10], but renal angiography remains the gold standard for diagnosing renal infarction. We suggest these two investigations should be performed before patients are diagnosed with loin pain haematuria syndrome. This also serves as a reminder that renal artery stenosis secondary to FMD is not always associated with hypertension.

We suggest that loin pain and microscopic haematuria should not be taken lightly and rare causes such as FMD should be included in the differential diagnosis.

Conflict of interest statement. None declared.

References

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