Fig. 21.10.2.2 Magnetic resonance angiogram demons

Fig. 21.10.2.2 Magnetic resonance angiogram demonstrating bilateralrenal artery stenosis (arrows) in a patient with Takayasu’s arteritis.

section 21 Disorders of the kidney and urinary tract 4992 ANCA-associated vasculitis Following a prodromal phase of several months with constitutional symptoms such as fever, night sweats, polymyalgia, and weight loss, patients present either with an extrarenal manifestation, or symptoms of constitutional disturbance, or evidence of nephritis. Nonrenal spe- cialties receiving vasculitis referrals, such as ear, nose, and throat medi- cine, need an awareness of the potential for systemic disease and access to ANCA testing and urgent nephrology referral (Box 21.10.2.1). Because renal vasculitis is asymptomatic until clinical features of renal insufficiency develop, the absence of specific symptoms of renal dis- ease often result in diagnostic delay. In consequence, patients with renal-limited vasculitis present with more advanced renal failure. The average delay from onset of symptoms to diagnosis has short- ened from longer than 1 year in the 1990s to less than 6 months. During this phase, urinary abnormalities will be present and should therefore be

sought in all patients with unexplained illness, or where there is a suspicion of vasculitis. Glomerular haematuria may be visible or nonvisible and is accompanied by subnephrotic proteinuria, but may be confused with prostatic disease or urinary tract infection. The pres- ence of red cell casts on urine microscopy reflects severe glomerular injury and is associated with crescentic glomerulonephritis. Atypical presentations including ‘failure to thrive’ and unexpected asymptom- atic renal impairment are more common in elderly patients. Renal vasculitis is more common in elderly patients with AAV, being present in 95%. Pulmonary involvement, typically with radio- logical infiltrates, reflects alveolar capillaritis in both GPA and MPA. The term pulmonary–renal syndrome is used to describe the clinical presentation of alveolar haemorrhage with crescentic glomerulo- nephritis. Other pulmonary features common in AAV include pul- monary fibrosis, bronchiectasis, and cavitating disease: these may precede or be detected at the same time as nephritis. Renal vasculitis commonly presents with the syndrome of rapidly progressive glomerulonephritis, that is, deteriorating renal function and crescentic glomerulonephritis on kidney biopsy. AAV accounts for 50 to 80% of cases of this syndrome and is differentiated from other causes by serological testing and renal immunofluorescence studies (Table 21.10.2.2). AAV with anti-GBM disease About 5% of patients with AAV present with simultaneous renal vasculitis and anti-GBM disease. They are older, have more severe renal disease, and are more likely to have pulmonary involvement than other AAV patients. The serology demonstrates ANCA posi- tivity, usually MPO-ANCA, and anti-GBM antibodies. Renal hist- ology reveals an aggressive crescentic glomerulonephritis, typically involving all glomeruli with linear IgG deposition on immunofluor- escence. When presenting in renal failure, such patients are more likely to recover renal function than in pure anti-GBM disease. However, after the initial presentation, unlike in anti-GBM disease, they can follow a relapsing course with persisting ANCA positivity. See Chapter 21.8.7 for further discussion of anti-GBM disease. IgA vasculitis (Henoch–Schönlein purpura) IgA vasculitis, although most frequent in children, occurs in adults, when it often pursues a relapsing course. By definition, extrarenal features of vasculitis including purpura, arthritis, and gastrointes- tinal involvement are present, but differentiation from other vascu- litic syndromes requires the demonstration of IgA deposition on skin or renal biopsy. A short prodromal period of 1 to 2 weeks with pur- pura often associated with a bacterial upper respiratory tract infec- tion are typical of IgA vasculitis. Purpura predominates in the lower limbs, then the upper limbs, then the trunk, and may coalesce and ulcerate. Some 50% of adults will have evidence of renal involvement, but it is important to recognize that this may be delayed by 2 to 4 weeks from the cutaneous presentation. Purpura is also common in cryoglobulinaemia, when digital ischaemia, livedo reticularis, neur- opathy, and other organ manifestations may also be seen. The renal presentation of IgA vasculitis overlaps with that of IgA nephropathy but is more likely to pursue a rapidly progressive course and to have extracapillary glomerular necrosis with crescents on biopsy. Polyarteritis nodosa Patients present with constitutional features of weight loss, fever, and night sweats, with specific symptoms depending on the ves- sels involved. Myalgia and muscle tenderness due to muscle in- volvement is common, as is abdominal pain due to intestinal ischaemia. Involvement of the skin can produce livedo reticularis, bullous/vesicular eruptions, and ulcers, and aneurysms can some- times form palpable nodules when they occur in subcutaneous tis- sues. Infarction of peripheral nerves presents with mononeuritis multiplex. Ischaemia in the kidney frequently manifests with hypertension: regional infarction—presenting with loin pain and haematuria—is rarely seen. Virtually any organ can be involved, with presentations ranging from myocardial ischaemia to orchitis. The association of polyarteritis nodosa with microscopic vessel involvement, such as necrotizing glomerulonephritis, has been called ‘polyangiitis overlap syndrome’, but is now classified as MPA. Takayasu’s arteritis Some 20%

of patients with Takayasu’s arteritis have renal arterial disease causing renal artery stenosis, hypertension, reduced renal size, and renal impairment (Fig. 21.10.2.2). See Chapter 19.11.6 for further discussion of Takayasu’s arteritis. Fig. 21.10.2.2 Magnetic resonance angiogram demonstrating bilateral renal artery stenosis (arrows) in a patient with Takayasu’s arteritis.

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