Headache and a syndrome of bilateral hydrenephrosis

Case

A 33-year-old woman presented in January 1980 with symptoms of persisting severe pain in the lower abdomen and weight loss. During menstruation there was worsening of the pain, nausea and vomiting. She had periods of headache, which impressed as a vascular headache. Seven years previously she consulted an internist because of general malaise. He found no cause; however the erythrocyte sedimentation rate (ESR) was 73 mm (1 h). During follow-up the ESR remained elevated.

By physical examination we found a blood pressure of 180/125 mmHg, but no other abnormalities. The ESR was 92 mm, renal and liver function tests were normal, serum IgG was 17.8 g/l (normal 6.8–11.2), serum IgA and IgM were normal. ANA and cryoglobulins were not found. Urinalysis showed no abnormalities, there was no proteinuria. By ultrasound there was a hydronephrosis on both sides. A CT scan of the abdomen showed enlarged lymph nodes in the vicinity of both kidneys and a solid process in the pelvis. The late phase of an angiography (Figure 1) showed proximal ureteral dilation caused by impression of a more distal process. An operation was performed.

Question

What is your diagnosis?

Fig. 1. Angiography (late phase): hydronephrosis, enlarged lymph nodes.
Answer to the quiz on the previous page

At surgery no signs of malignancy were found. Next to both tubae and the ureter an infiltrate and pathological lymph nodes was found. Microscopy showed a granulomatous vasculitis with giant cells in the periuretral and peritubular tissue (Figures 2 and 3). The patient was treated with corticosteroids, subsequently her complaints disappeared and the ESR normalized. The hydronephrosis disappeared (Figure 4). Blood pressure was treated successfully. At present she still needs antihypertensive drugs. Moreover, it was not possible to stop steroids, the dosage being 5 mg prednisolone od.

Although not entirely typical, the histological picture most closely resembled that of giant-cell arteritis (GCA). Vasculitis causing ureteral stenosis has been mentioned in the literature only in association with systemic diseases such as Wegener granulomatosis, polyarteritis nodosa, allergic granulomatosis and angitis, or drug-related vasculitis [1]. The vasculitis in our patient seems to be unique since GCA of the ureteral blood supply has never been described before. The systemic spread of GCA has been known for 50 years [2]. The majority of cases of GCA present in a definable clinical syndrome [3]. Frequent manifestations include fatigue, headache, jaw claudication, loss of vision, scalp tenderness, polymyalgia rheumatica and aortic arch syndrome. However, our case shows that the clinical picture can be much less specific. Gynaecological involvement in GCA has been described, even in the absence of typical clinical findings such as temporal arteritis [4,5]. Often this diagnosis is made after surgery for suspected malignancy or at the end of a complete check-up for fever of unknown origin, as in our case. Ganesan et al. [5], studied forty-six cases of vasculitis affecting the female genital tract; GCA was present in four cases. Only one of them had a proven GCA on a temporal biopsy. None of the studied cases had evidence of ureteral involvement.

The aetiology of GCA is unknown. Reports of its presence in first-degree relatives of GCA patients, a predilection for the disease to occur in caucasians, and an association with HLA-DR4 suggest a genetic predisposition. Variations of incidence in different areas suggest that, in addition to genetic predisposition, environmental factors may be important. Furthermore, ageing seems to be an important factor as GCA is seldom diagnosed below the age of 50 years. Treatment of GCA with corticosteroids is highly effective. With an adequate dose of prednisolone in...
the first month good symptomatic control and a fall in ESR will be obtained. Subsequently one must aim for maintenance doses of less than 10 mg per day after 6 months. The exact dose requires an adjustment to the needs of the individual patient. GCA tends to run a self-limited course, lasting from several months to several years [6]. Exacerbations and recurrences are seen in some patients.

In summary, we present a patient with a rather non-specific presentation of GCA. This case shows that one should consider this diagnosis as a cause of unclear ureteral stenosis.

References


Ilanit Z. Hene1
Roel Goldschmeding2
Ronald J. Hene1

1Department of Nephrology
2Department of Pathology
University Medical Centre
Utrecht
The Netherlands
Email: r.j.hene@azu.nl