Letter and Replies

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Nutcracker phenomenon or nutcracker syndrome?

Sir,

Chang et al. [1] describe a young patient in whom a nutcracker syndrome was diagnosed by magnetic resonance imaging and magnetic resonance angiography. Patients with nutcracker phenomenon are usually young and slim, and the diagnosis can be made by ultrasound. There are established criteria for the diagnosis of the entrapment of the left renal vein by two-dimensional ultrasonography and Doppler sonography [2,3].

Nutcracker phenomenon, an entrapment of the left renal vein usually in the fork between the aorta and the superior mesenteric artery, was first described by de Schepper in 1972 [4]. Differentiation should be made between an asymptomatic dilatation of the left renal vein (‘nutcracker phenomenon’) and the ‘nutcracker syndrome’ presenting with gross or microscopic haematuria, varicoceles and hypertension. The nutcracker syndrome is said to be an underdiagnosed cause of haematuria [5]. In 52 children with microscopic haematuria, one-third fulfilled the ultrasonographic criteria for nutcracker phenomenon [2]. However, when the left vein is routinely examined by ultrasound and/or duplex, the nutcracker phenomenon is quite frequently seen and nutcracker syndrome might become an overdiagnosed entity, with the danger that the real causes of haematuria are not recognized. We have seen a 12-year-old girl with microhaematuria. She had a massive dilated left renal vein. Renal biopsy, however, had revealed IgA nephropathy. The nutcracker phenomenon in combination with IgA nephropathy has been described in other cases [6]. Therefore, it seems to be wise not to forget the differential diagnosis if a dilated left renal vein is seen in a patient with haematuria or (orthostatic) proteinuria.

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Reply

Sir,

Liebl cites an example of the co-existence of IgA nephropathy and nutcracker phenomenon, diagnosed in Germany. We always feel that the correct diagnosis of nutcracker syndrome is possible only after a detailed and thorough consideration of clinical history as well as physical and laboratory findings. Furthermore, we never underestimate the wide variability and unpredictability of medical practice, either in Europe or in Asia. In this regard, we agree that a radiographic demonstration of a dilated left renal vein is not equivalent to nutcracker syndrome, nor does the pathological demonstration of renal mesangial IgA and C3 deposits necessarily equate to primary IgA nephropathy. The deposits might be secondary to a systemic disease such as polymyositis/dermatomyositis, as we reported previously [1,2]. What we are trying to emphasize is that clinicians must always look for the primary cause of the disease, rather than making a laboratory-based diagnosis, which could be misleading.

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