Transient arthritis with positive tests for rheumatoid factor as presenting sign of shunt nephritis

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Abstract

Shunt nephritis is a rare complication of a chronically infected ventriculoatrial shunt. A 17 year old boy is described, with arthritis in both ankles and positive rheumatoid factor tests, who presented with symptoms of shunt nephritis. Blood cultures were positive for *Staphylococcus epidermidis*. The patient recovered completely after removal of the shunt and antibiotic treatment. Shunt nephritis should be considered in patients with a ventriculoatrial shunt presenting with arthritis, and the arthritis may be an early indicator of an infected ventriculoatrial shunt.

Since the first description in 1965,¹ several reports have been published on the so-called 'shunt nephritis' as a consequence of a chronically infected ventriculoatrial shunt.²⁻³ Immune complex formation is considered to underly the pathophysiology of this disease.²⁻³ Arthritis as a manifestation of shunt nephritis has been reported only incidentally.⁴⁻⁵ We report an additional case of a 17 year old boy with shunt nephritis, who presented with arthritis of both ankles and positive rheumatoid factor tests.

Case report

The previous history of the patient included implantation of a ventriculoatrial shunt because of hydrocephalus in June 1985. In August 1985 fever and a wound infection with *Staphylococcus epidermidis* at the site of implantation were treated with antibiotics. Eight months later the patient was referred to a local hospital because of arthritis in both ankles of two weeks' duration and purpura on the lower legs. The arthritis subsided after one week without treatment and a joint puncture was not performed. The Waaler-Rose (1/512) and latex fixation tests ($\geq 1/10\ 000$) were strongly positive. In addition, a raised erythrocyte sedimentation rate, decreased complement C3 concentrations, proteinuria, and microhaematuria were found. The patient was referred to our hospital for further evaluation in October 1986. His history included recurrent episodes of purpura on the lower legs, but arthritis or arthralgia had not recurred and antibiotics had not been used for one year.

Physical examination disclosed purpura on the extensor sides of the lower legs but was otherwise normal. Laboratory tests showed the following: erythrocyte sedimentation rate 100 mm in the first hour and C reactive protein 96 mg/l (normal <6). Haemoglobin 100 g/l, leuco-

cytes $5.3 \times 10^9/l$, platelets $222 \times 10^9/l$. Serum creatinine was 84 $\mu\text{mol/l}$ (normal <106), serum urea 5.7 mmol/l (normal <6.7). The glomerular filtration rate was impaired (86 ml/min), proteinuria amounted to 0.7 gram protein per day, and the urinary sediment contained 5-10 erythrocytes (high power field) without casts. IgG was 42.6 g/l (normal 8.5-15.0), IgM 2.9 g/l (normal 0.6-2.6), IgA 2.3 g/l (normal 0.9-4.5), complement C3 1.09 g/l (normal 0.64-1.20), and C4 0.08 g/l (normal 0.11-0.40). A test for circulating immune complexes was strongly positive (indirect granulocyte phagocytosis test⁶), but tests for cryoglobulins were not performed. Antinuclear and anti-double-stranded DNA antibodies could not be detected. The IgM rheumatoid factor level (enzyme linked immunosorbent assay (ELISA)) was 170 IU/ml (normal <10). A biopsy specimen from a purpura lesion disclosed leucocytoclastic vasculitis (fig 1) with granular deposition of IgM and C3c at the dermoepidermal junction, and IgM, C3c, and fibrinogen in the vessel walls. Unfortunately, a renal biopsy specimen yielded no glomeruli. The echocardiograph showed no abnormalities. Blood cultures grew *Staphylococ-*

Figure 1 (A) Classical picture of leucocytoclastic vasculitis. Small arterioles and venules surrounded by an infiltrate containing many neutrophils and nuclear dust with fibrinoid necrosis of vessel walls and extravasation of erythrocytes. (B) The same specimen at greater magnification. (Haematoxylin and eosin.)

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Figure 2 Clinical course in relation to laboratory measurements in this patient. V-A shunt=ventriculoatrial shunt.

cus epidermidis with a pattern of resistance similar to that from the wound infection in August 1985. *Staphylococcus epidermidis* was found in the ventriculoatrial shunt, which was subsequently removed, and oral antibiotics were given for two weeks. The patient recovered completely with the laboratory tests returning to normal (fig 2).

Discussion

A patient is described with shunt nephritis presenting with transient arthritis in both ankles and positive rheumatoid factor tests. Although infection of a ventriculoatrial shunt often occurs (27%), it is rarely followed by a shunt nephritis (4%).⁷ Since the original description in 1965¹ more than 70 cases of shunt nephritis have been described.² Non-pathogenic bacteria, usually *Staphylococcus epidermidis* (75%) as in this patient, are the most common infecting organisms,² and the clinical manifestations of shunt nephritis include fever, purpura, and signs of renal disease. Renal complications generally predominate, and a biopsy will usually show mesangiocapillary or diffuse proliferative glomerulonephritis.² This patient had clinical symptoms compatible with mild glomerulonephritis, but unfortunately no glomeruli were to be seen in the renal biopsy specimen. Shunt nephritis is assumed to be an immune complex disease^{2,3} with manifestations resembling those of subacute bacterial endocarditis. Serum antibodies to the offending microbe have been detected in shunt nephritis,³ and deposition of bacterial antigen has been shown in the glomeruli.⁸ Also, cryoprecipitates

have been shown to contain bacterial antigen or antibody in some cases.⁹ Evidence in favour of the immune complex nature of shunt nephritis in our patient is the strongly positive test for circulating immune complexes, the decreased concentrations of C3 and C4, and the immunofluorescence findings in the skin biopsy specimen.

As in subacute bacterial endocarditis, positive rheumatoid factor tests also can be found in shunt nephritis,² whereas in contrast with subacute bacterial endocarditis, arthritis has been reported only rarely^{4,5} and has been designated 'shunt arthritis' in this condition.⁴ It may be due to the deposition of circulating immune complexes in the synovium as has been shown in subacute bacterial endocarditis.¹⁰ Although a joint puncture was not performed in this patient, the disappearance of arthritis without antibiotic treatment argues against septic arthritis and is in favour of 'shunt arthritis'. Shunt nephritis should be considered in patients with a ventriculoatrial shunt presenting with arthritis, and this arthritis may be an early indicator of an infected ventriculoatrial shunt in such patients. Multiple blood cultures, which are positive in 85% of the cases,³ have to be performed even in the absence of fever. Antibiotic treatment alone is generally not sufficient, and removal of the ventriculoatrial shunt is nearly always necessary to obtain complete recovery.^{2,7} This patient fully recovered with a complete disappearance of urinary abnormalities after removal of the ventriculoatrial shunt and antibiotic treatment.

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