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A Case of Shunt Nephritis

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Nephritis associated with a chronically infected ventriculoatrial shunt is known as shunt nephritis. A 6-year-old girl with prior history of a ventriculoatrial shunt presented complaining of fever and gross hematuria. Serum complement levels were decreased and a coagulase-negative S. epidermidis was cultured from her blood. The renal biopsy specimen showed features of membranoproliferative glomerluonephritis type I. Hydrocephalus was so severe that shunt removal was impossible. With antibiotic therapy, clinical symptoms and laboratory findings include complement levels were normalized although microscopic hematuria persisted. To our knowledge, this is the first case of shunt nephritis in Korea. In addition to the case report, a brief review of shunt nephritis has been added.

Key Words : *Shunt infection, Staphylococcus epidermidis, Glomerulonephritis.*

INTRODUCTION

The overall incidence of ventricular shunt infection varies between 3% and 11% (Narchi et al., 1988). It is a significant cause of death in these patients, who may present in one of four ways : 1) with sepsis ; 2) with signs of increased intracranial pressure ; 3) as an asymptomatic state with positive shunt CSF cultures associated with leukocytosis and hypoglycorrhachia ; or 4) with renal disease (Black et al., 1965 ; Kaufman and Mcintosh, 1971). The development of nephritis as a complication of chronic bacteremia in patients with hydrocephalus and infected ventriculoatrial shunts has been reported on numerous previous occasions. The condition develops as a result of the formation of immune complexes, which activate the complement system and deposit in the renal glomeruli(Dobrin et al., 1975). Treatment of the disorder, which requires removal of the infected shunt and long term administration of antibiotics, however, can cause irreversible renal parenchymal damage. Recently, We have encountered a case of nephritis associated with Staphylococcus epidermidis infection in a ventriculoatrial shunt. To our knowledge, this is the first case report of its kind in Korea.

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CASE REPORT

A 6-year-old girl was hospitalized for the chief complaints of intermittent high fever and gross hematuria which she had had for 1 month. A ventriculoperitoneal shunt had been performed when she was 6 months old because of noncommunicating hydrocephalus. Over the next 2 years, the shunt became obstructed and was revised three times. This shunt was replaced with a ventriculoatrial shunt when she was 3 years old. Physical examination on admission revealed an alert consciousness, pale appearance and normotensive child in no acute distress. Blood pressure was 90/60 mmHg and

temperature was 38℃. Her liver edge was palpable 3 cm below the right costal margin and her spleen tip 3 cm below the left costal margin. The remainder of the physical examination was unremarkable. Laboratory studies disclosed the following values : hemoglobin 8 gm/dl, WBC 7,300/mm^3 with a normal differential count, ESR 46 mm/1hr, blood urea nitrogen 17 mg/dl, creatinine 0.6 mg/dl, serum albumin 2.8 gm/dl, serum cholesterol 156 mg/dl, CRP 4+, RA+, ASO 200 IU/ml, C_3 21 mg/dl, C_4 4mg/dl, cryoglobulin-, circulating immune complex-. Urinalysis disclosed 3.5 gm/day of protein, many RBC/hpf, WBC/hpf and some granular cast. Blood culture grew a coagulase-negative S. epidermidis. Intravenous cefazolin was given from the 2nd day. Two days after the start of antibiotics, there was no high fever but she continued to have mild fever. At 5 days, she again developed high fever and a coagulase-negative S. epidermidis was isolated from a 2nd blood culture in spite of sensitivity to cefazolin. At 7 days, cefazolin was discontinued and teicoplanin begun. Following intravenous teicoplanin therapy for 4wks, the hepatosplenomegaly had resolved, the bacteremia cleared, serum complement levels returned to normal and urinalysis was normal except for microscopic hematuria.

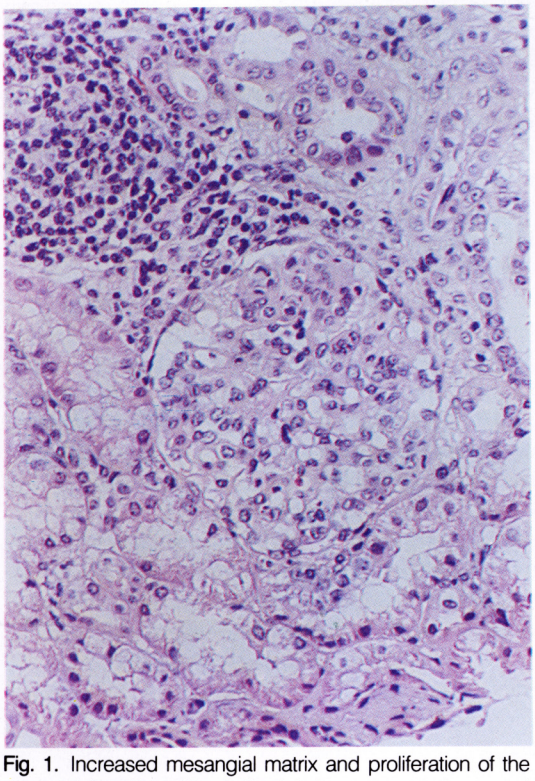

Fig. 1. Increased mesangial matrix and proliferation of the mesangial cells with slight lobular accentuations and some neutrophilic infiltrations are seen. Interstitium shows folliculoid lymphocytic infiltrations(H-E stain).

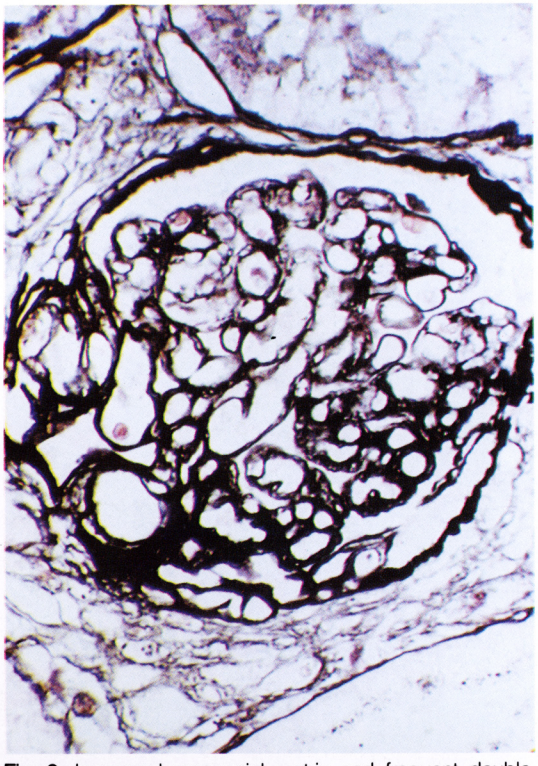

Fig. 2. Increased mesangial matrix and frequent double track appearanace of the basement membranes are seen-(PA-Silver stain).

A percutaneous renal biopsy was performed on the 14th day. Light microscopy revealed diffuse increase of the mesangial matrix and minimal proliferation of mesangial cells with occasional neutrophilic infiltration and double track appearance of the basement membranes(Fig. 1) (Fig. 2). Capillary walls are segmentally thickening. Interstitium shows patch lymphocytic infiltration and tubules show some acidophilic cast like material with atrophic change. Ultrastructural study is performed on one glomerulus, showing global increase of the mesangial matrix, thickenings of the peripheral capillary walls with frequent mesangial interposition and subendothelial electron dense de-

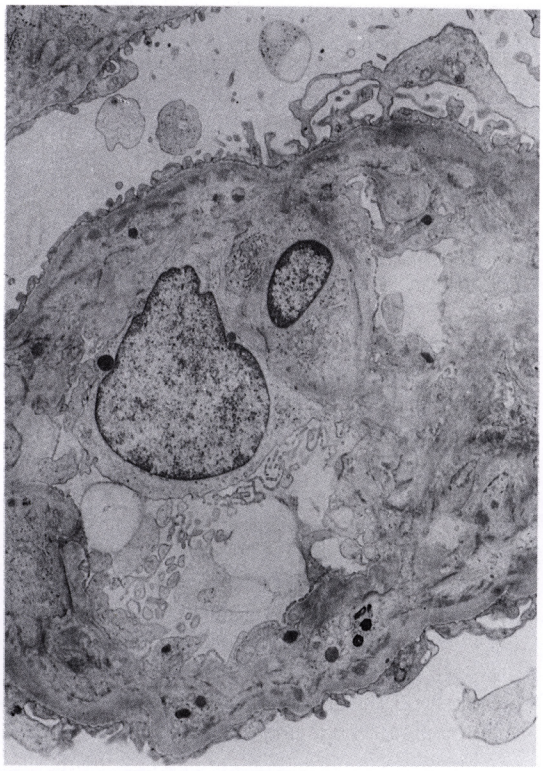

Fig. 3. Mesangial interpositions with subendothelial electron dense deposits and obliteration of the epithelial foot processes are seen. Note a neutrophil adhered to the endothelial cell surface(EM,×6,000)

posits(Fig. 3). Capillary lumens are partly obliterated and the epithelial foot processes are segmentally effaced. Several neutrophilic marginations are also noted. The biopsy specimen was insufficient for immunofluorescent studies. She was discharged and has been followed as an out patient. 14 months later, she did not display fever, hepatosplenomegaly or anemia but serum complement levels were slightly decreased and microscopic hematuria persisted.

DISCUSSION

Shunt nephritis was first described in 1965 (Black et al., 1965). Since then, it has been found to occur in 0.7% to 2.25% of patients with an infected shunt and, in total, less than 86 cases have been reported in the literature (Narchi et al., 1988). Symptoms appear in a well-defined pattern (Black et al., 1965 ; Rames et al., 1970). A review of the 86 published cases gave the following presentation and incidences : hematuria(89%), fever(88%), anemia(86%), proteinuria(70%), hepatosplenomegaly(55%), nephrotic syndrome(30%), nonthrombocytopenic purpura-(19%) and hypertension(15%) (Narchi et al., 1988). In our case, there was fever, anemia, hematuria, proteinuria, hepatosplenomegaly but nephrotic syndrome, purpura and hypertension were absent. Other symptoms associated with VA shunting include urticaria vasculitis, necrotizing cutaneous vasculitis and arthritis. An outstanding characteristic of the disease is the long interval from shunt placement to recognition of symptoms, averaging about 3 years.

Laboratory findings are characteristic (Kaufman and Mcintosh, 1971 ; Wald and Mclaurin, 1978). Urinalysis reveals proteinuria and microscopic or occult hematuria. An iron deficiency anemia or a moderate normochromic normocytic anemia is usually present, probably due to decreased erythropoietin production. Renal function may vary from mild azotemia to temporary oliguria and even to anuria. Coagulase negative S. epidermidis has been reported in over 70% of these patients (Rames et al., 1970 ; Narchi et al., 1988). Other organisms such as S. aureus, Streptococcus, Propionibacterium acnes, Listeria monocytogenes, Corynebacterium bovis, Diphtheroids, Micrococcus, Pseudomonas aeruginosa, Cryptococcus neoformans, and Filamentous histoplasma capsulatum have also been described in cases of infected VA, and ventriculojugular shunts (Arze et al., 1983). In our case, coagulase negative S. epidermidis was also isolated from blood culture. Immunological studies have suggested that shunt nephritis is analogous to other known immune complex diseases of the kidney, including those associated with subacute bacterial endocarditis, syphilis, osteomyelitis, and some viral infections (Dobirin et al., 1975). Laboratory analyses have shown low serum C_3 concentrations not necessarily corresponding to the severity of the disease, the presence of cryoglobulins, and a positive rheumatoid factor test, all of which were reversible when the shunt was completely removed (Beeler et al., 1976 ; Wakabayashi et al., 1985). The return of depressed serum C3 and C4 protein complement to normal levels coincides with the recovery of infection and clinical remission of the glomerulonephritis. Our case on admission disclosed low serum complement and a positive rheumatoid factor but cryoglobulins were negative. Following antibiotic therapy for

2 weeks, the complement returned to normal. Renal tissue biopsies show characteristic endothelial proliferation with mesangial expansion and proliliferation of cellular elements. Hyperlobulated, tufted, and enlarged glomeruli with a reduced number of open capillary lumina are seen (Arze et al., 1983 ; Wakabayashi et al., 1985). Subendothelial electron dense deposits are noted by electron microscopy. Immunofluorescence studies showed granular IgG, IgM, IgA, C_3, C_4 and C_{1q} deposits along the capillary wall (Arze et al., 1983 ; Narchi et al., 1988). Also in our case, renal biopsy showed increased mesangial matrix and mesangial interpositions with subendothelial electron dense deposits noted by electron microscopy.

The cornerstone of therapy is the elimination of the infection. Regarding the therapy for shunt infection, the removal of the entire foreign body and concomitant systemic administration of specific antibiotics has been recommended (Shurtleff et al., 1971), giving the best chances for cure, although 50% of cases could be managed without surgical intervention. In view of renal disease, however, the first policy is to be prefered and has been followed most often, while removal only can be also successful (Harkiss et al., 1979). In our case, hydrocephalus was so severe that shunt removal was not done. Antibiotics were given instead, which resulted in much improvement of clinical and laboratory findings including serum complements and nephrotic range proteinuria, however, the complement levels were slightly decreased and microscopic hematuria persisted during the follow up period of 14 months. We think that decreased serum complement levels and persisted microscopic hematuria were due to a remainded shunt which acted as a foreign body. So, the treatment of shunt nephritis was maybe incomplete. Monthly careful follow up of urinalysis and laboratory studies are mandatory in shunted patients, since early diagnosis and treatment is the best way to prevent irreversible renal damage.

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