Lupus Cystitis in the Japanese

Lupus cystitis is an uncommon but definite manifestation of systemic lupus erythematosus (SLE). Interstitial cystitis, which in itself is also an uncommon lesion of the urinary bladder was apparently first described by Nitze (1) in 1907. Hunner (2) considered it to be a rare type of bladder ulcer. Patients with this form of SLE have infrapubic pain which diminishes with voiding. Urinalysis reveals either no abnormality or only microscopic hematuria and is characterized by a negative cytology. A patient with interstitial cystitis complicated with SLE was reported by Fister (3) in 1938. Shipton (4) also reported a case of chronic interstitial cystitis as Hunner's ulcer with SLE. Interstitial cystitis complicated with SLE was termed "lupus cystitis" by Orth et al in 1983 (5). They described six patients who had a reduced bladder capacity and a thickened and irregular bladder wall as seen on intravenous urography. Some patients had a hydronephrosis and hydroureters because of obstruction of the ureterovesical junction. Five of the six patients had gastrointestinal involvement and two had central nervous system (CNS) involvement. Although Orth et al (5) defined the clinical entity of lupus cystitis several years ago, there are few reports of such cases and an accurate definition of lupus cystitis has yet to be clearly established.

In Japan, the first case of lupus cystitis was reported in 1984 (6). Twenty-seven cases of lupus cystitis have since been reported and the clinical entity of lupus cystitis has become much better understood. All cases satisfied the ARA SLE criteria revised in 1982 and the diagnoses can be made by biopsy. Pollakisuria without abnormality in urinalysis was evident in all these cases. Most reports stated a volume reduction in the urinary bladder and hydronephrosis was noted on intravenous pyelography. An accurate diagnosis is difficult and varies considerably from typical bacterial cystitis. Without adequate therapy, hydronephrosis can lead to renal failure and the necessity of nephrostomy due to the irreversible diminished capacity of the bladder.

See also p. 155

Interstitial cystitis is not coincidentally complicated in SLE because SLE plus this condition has particular clinical features. All but one of 27 patients had symptoms of diarrhea, nausea and vomiting. Furthermore, in more than half, gastrointestinal symptoms preceded symptoms of interstitial cystitis. In some advanced cases, protein-losing enteropathy, paralytic ileus or gastrointestinal hemorrhage occurred. Moriuchi et al (7) had the first case report of in a Japanese patient published in English; this patient’s bowel had perforated. Therefore, the main lesion might reside in the gastrointestinal tract, or both symptoms might have derived from the same etiology.

Segawa et al (8) reported the second case of lupus cystitis in Japan; the patient had no gastrointestinal symptoms nor was the CNS involved. Orth et al (5) stated that involvement of the CNS is a typical characteristic of this condition. However, based on the results of the 27 cases reported from Japan, there appears to be no significant relationship between lupus cystitis and CNS involvement. Thus, the prevalence of major organ involvement, including lupus nephritis, is not significantly higher than that of general SLE cases.

The prevalence of interstitial cystitis is estimated to be 10.6 of 100,000, of which 92% of the patients are women (9). Interstitial cystitis is accompanied not only by SLE but also by collagen diseases such as Sjögren’s syndrome (10) or rheumatoid arthritis. As for the incidence of lupus cystitis, Alarcon-Segovia et al (11) investigated autopsies of SLE patients and noted interstitial cystitis in 11 of 35 autopsies. However, the manifestations are estimated to be 0.5–1% of all SLE cases.

The precise mechanisms involved in lupus cystitis remain obscure. Immune complex-mediated vasculitis may have an important role because deposition of the immune complex containing IgG; C3c was demonstrated along the small articules in the bladder of some patients (12, 13), while involvement of various cytokines in lupus cystitis is also not clear. Segawa et al (8) noted the elevation of interleukin-8 (IL-8) and monocyte chemotactic and activating factor (MCAF), as urinary chemokines, and their resolution after treatment. These findings provide insight to the mechanism of lupus cystitis. In terms of autoantibodies, there seems to be no significant correlation between lupus cystitis and autoantibodies. In one study, anti-intermediate filament antibodies were considered to be specific antibodies to this disease, due to the involvement of intestine and bladder (14).

According to a meta-analysis of data in reports from Japan, the prognosis of urinary bladder function in patients with lupus cystitis was closely related to the period from the appearance of the symptoms to the initiation of steroid therapy. In most in the poor prognosis group, steroid therapy was initiated more than six months after the appearance of cystitis and the dose of steroid was not related to the prognosis. In contrast, the patient reported in the present issue (8) was effectively treated with steroid pulse therapy 4 years after the onset of cystitis.

While the importance of an early diagnosis and initiation of steroid therapy has been stressed, the diagnosis of this entity may not have always been accurate. General practitioners and urologists should completely understand the factors related to
lupus cystitis in order to effectively treat such patients.

Takao KOIKE, MD and Katsuhiko TAKABAYASHI*, MD
Department of Medicine II, Hokkaido University School of Medicine, Sapporo 060
*Department of Medicine II, Chiba University School of Medicine, Chiba 260

References