Idiopathic Retroperitoneal Fibrosis and HLA Antigens

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Dear Sir,

The etiology of retroperitoneal fibrosis (RPF), or Ormond’s disease, is still unknown. Injury, infection, drugs and familial factors have been suggested, but in most cases no real cause has been found. Local immune mediation around the periaortic wall inducing the proliferation of fibroblasts and collagen fibrils has been hypothesized [1], which may account for the efficiency of empiric corticotherapy. It has been associated with class I HLA-B27 in 5 out of 7 published reports [1-7] of HLA determination. Thus, HLA-B27 could suggest a possible genetic factor in this benign neoplasm, and may be a helpful marker of the disease.

The results for class I and II HLA antigens from 8 men are summarized in table 1. The patients have been treated in two nephrology units for the last 17 years. Their ages range from 28 to 65 years (mean 46). None of the known causes of this disease were observed, but abdominal injury had occurred before diagnosis in 3 cases, and the oldest patient (tumor involving renal arteries) had received antihypertensive therapy. Steroids were started in 7 cases, after RPF had been diagnosed during surgery and the pathological criteria had been fulfilled. All but one of the patients are still alive and therapy has been withdrawn (mean duration: 17 months). Subsequent CT scans of 5 of the subjects have shown incomplete but stable regression of RPF. The last patient (No. 8) had a biopsy for diagnostic procedures and is still being treated with tamoxifen. None of these patients had an immunologic abnormality in the serum.

Our systematic screening carried out on Caucasian RPF patients has not revealed an HLA-associated marker (patients vs. blood bank donor controls; data not shown). Moreover, it does not confirm that HLA-B27 is linked to the disease. It has to be highlighted that 3 of 5 patients with HLA-B27 previously diagnosed as having RPF developed chronic sacroiliitis in association with fibrosis.

References