Introduction—Hepatitis C virus infection (HCV) is the main cause of mixed cryoglobulinemia vasculitis (CryoVas). Data on infectious mixed CryoVas in the absence of HCV infection are lacking. We aimed to analyze the features of patients with infectious mixed CryoVas in the absence of HCV infection included in the French CryoVas survey.

Methods—We have included 260 patients with non-HCV mixed CryoVas diagnosed between January, 1995 and July, 2010. Among them, 18 patients presented with infectious mixed CryoVas. Demographical, clinical and biological data, as well as therapy and outcome, were assessed.

Results—Infectious causes were: virus infection in eight patients [hepatitis B virus (HBV) in four, and CMV, EBV, parvovirus B19 and HIV in one case each], pyogenic bacterial infection in six patients, parasitic infection in two patients, and leprosy and candidiasis in one case each.

Baseline manifestations were: purpura (78%), glomerulonephritis (28%), arthralgia/arthritis (28%), peripheral neuropathy (22%), necrosis (22%), cutaneous ulcers (17%), and myalgia (11%). Cryoglobulinemia was type II in 12 patients (67%) and type III in six (33%).

As first-line therapy, six patients received corticosteroids, one cyclophosphamide and none rituximab, but 14 patients received anti-infectious specific therapy. Among the latter, ten were in sustained remission of the disease, two died of the underlying infectious disease, and two had refractory or relapsing disease related to HBV infection treated with rituximab in addition to antiviral therapy, leading to complete remission. The four remaining patients who did not receive specific therapy had CMV, EBV, parvovirus B19 and HBV infection, and remained in remission of the CryoVas.

Conclusion—In the absence of HCV infection, virus and pyogenic bacterial infections represent the main causes of infectious mixed CryoVas. Anti-infectious therapy is frequently associated with remission. Immunosuppressive agents should be considered only in patients with refractory and/or life-threatening vasculitis.

http://dx.doi.org/10.1016/j.lpm.2013.02.101