Characteristics of patients with infectious cryoglobulinemia vasculitis in the absence of HCV infection: Results from the French nationwide CryoVas survey

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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%). Cryoglobulinaemia 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 therapy. Among the latter, ten were in sustained 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.

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Ribavirin/protease inhibitor combination in hepatitis C virus associated mixed cryoglobulinemia vasculitis/Peg-INFα/

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Introduction.—The standard of care treatment of patients with hepatitis C virus (HCV)-mixed cryoglobulinemia (MC) vasculitis includes pegylated interferon alpha (PegIFNα)-alpha plus ribavirin and/or rituximab. Thirty to 40% of patients are non-responders or relapsers to such combination.

Objective.—To analyze the safety and efficacy of Peg-IFNa/ribavirin/Protease inhibitor combination in HCV-MC vasculitis.

Patients and methods.—Open label prospective cohort study including 23 patients with HCV-MC vasculitis. Peg-IFNa/ribavirin was associated to telaprevir [375 mg three times daily, for 12 weeks, (n = 13)] or boceprevir [800 mg three times daily, for 44 weeks, (n = 8)] for 48 weeks.

Results.—The median age was 59 (52.5–66) years, with 48.8% of women. Thirteen patients (56.5%) were complete clinical responders, and ten (43.5%) were partial responders at week 24. The virological response (i.e. HCV RNA negativation) was of 69.6% at week 24 (P = 0.005). The cryoglobulin level decreased from 0.44 to 0.06 g/L (P = 0.0006) and the C4 level increased from 0.09 to 0.15 g/L (P = 0.045). Grade 3 and 4 adverse events (mainly anemia, neutropenia and thrombocytopenia) were observed in ten cases (43.5%). Twenty patients (87%) received erythropoetin, nine (39.1%) had red cell transfusion and 2 (8.7%) had granulocyte stimulating agents. Antiviral therapy discontinuation was required in 8 (34.7%) patients for virological non response (n = 5), virological relapse (n = 2) and depression (n = 1).

Conclusion.—Peg-INFα/ribavirin/protease inhibitor combination seems highly effective in HCV-MC. Such therapeutic regimen should be administered cautiously considering the high rate of side effects.

Further readings