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Cardiac Sarcoidosis

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Description

Sarcoidosis is a systemic granulomatosis of unknown etiology, although many genetic and environmental factors have been suggested in its pathophysiology. The most commonly affected organs are the lung, lymphatic system, skin and eyes [1]. Symptomatic cardiac involvement is reported in 5 to 11% of cases. Asymptomatic cardiac involvement appears to be much more frequent, estimated to be up to 25% in autopsy series [2,3]. Cardiac involvement can be indicative of sarcoidosis. It has been reported that 16-35% of patients with complete atrioventricular block or ventricular tachycardia of unknown etiology will be diagnosed with cardiac sarcoidosis [4]. The main clinical presentations of cardiac involvement from sarcoidosis range from manifestations of heart failure to … sudden death related to a cardiac conduction or rhythm disorder. The other presentations are much rarer: atrial arrhythmia, valve disease, myocarditis, coronary vasculitis or even arrhythmogenic cardiomyopathy of the right ventricle.

Prognostic Factors

Cardiac involvement from sarcoidosis negatively influences the prognosis, accounting for 25% of the causes of sarcoidosis death in the United States and up to 85% in the Japanese series. Significant left ventricular dysfunction (ejection fraction less than 30%) is a major predictor since it is associated with one-year survival of 91%, five-year survival of 57% and ten-year survival of 19% [5,6]. Several studies have reported a good prognosis, i.e. the absence of a cardiac event after prolonged follow-up, in patients with clinically asymptomatic cardiac involvement, although these results remain debatable. In a recent French study, involving a large cohort of 157 patients with heart disease from sarcoidosis (according to the new diagnostic criteria) [7], ten-year survival rate was good (90%) [8]. Deaths were associated with age, low left ventricular ejection fraction, and the presence of late hypersignals on cardiac MRI. The contribution of PET scans to analyze the extent and activity of the disease, as well as the cardiac response to treatments, has been suggested by several recent studies. Technologies combining PET scans and MRI make it possible to differentiate active inflammatory lesions from sequelae fibrosing lesions [9-11]. If the vital prognosis of patients with cardiac sarcoidosis is relatively good, morbidity remains high due to the numerous relapses. The factors associated with these relapses are the presence of left heart failure and ventricular wall abnormalities on ultrasound [1,3,7].

Therapeutic Strategy

Old, non-randomized studies have suggested that systemic corticosteroid therapy should be offered as soon as possible because it has good efficacy on ventricular arrhythmias, signs of acute heart failure and atrioventricular conduction disorders. However, no noticeable difference was noted between high (>40 mg/day) or low doses (<30 mg/day) of corticosteroids. The place of immunosuppressive treatments has until now been reserved, as second-line treatments, for refractory cases and/or in cases of poor tolerance of corticosteroids [12]. Several drugs have shown some benefit, including methotrexate, cyclophosphamide, mycophenolate mofetyl and more recently infliximab [13-15]. Certain cardiological treatments may also be essential, in particular a pacemaker in the event of severe atrioventricular conduction disorders, or an implantable defibrillator in the event of severe rhythmic disorders. In a recent study on a large cohort of patients with cardiac involvement from sarcoidosis, after a median follow-up of 7 years, only intravenous cyclophosphamide was associated with a significant decrease in cardiac relapses. Other immunosuppressants (corticosteroids alone, methotrexate or mycophenolate mofetyl) were also associated with a lower risk of cardiac relapse but not statistically significant, possibly due to lack of potency [8]. Of note, several recent publications have reported encouraging results with infliximab in this indication, particularly in forms that are multi-refractory to conventional immunosuppressants [16-18].

The spontaneous prognosis of cardiac attacks from sarcoidosis justifies an "aggressive" therapeutic strategy comprising corticosteroid therapy and immunosuppressive treatment. Future prospective studies should focus on showing which of the corticosteroid/immunosuppressant combinations is the best performing and best tolerated, including by integrating the described unfavorable prognostic factors into the stratification.

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