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Perspective

The Diagnosis and Management of IgG4-Related Pericarditis

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INTRODUCTION

IgG4 related pericarditis has rarely been reported. The diagnosis of IgG4 related pericarditis is often difficult in terms of histologically analysis. PET/CT help identify IgG4 related organs and is useful to evaluate the efficacy of steroid therapy for IgG4 related disease. Most patients with IgG4-related pericarditis show a rapid clinical response to steroid therapy and it possibly prevents the progressive involvement of other organs. Prompt treatment contributes to improve the status of the disease.

DESCRIPTION

IgG4 Realated Disease (IgG4-RD) is a relatively rare autoimmune disorder associated with elevated serum IgG4 concentrations, lymphoplasmacytic infiltrate of IgG4 positive plasma cells and storiform fibrosis in various organs. The disorder was first reported in 2001 in patients with autoimmune pancreatitis. The involvement in pericardium is rare but a growing number of cases have been reported. Doumen, et al. summarized cases of IgG4-RD with pericardial involvement. In total of 33 case, the sole manifestation of IgG4 pericarditis is small number.

The comprehensive diagnostic criteria for IgG4-RD revised in 2020 proposed following conditions: 1) One or more organs show diffuse or localized swelling or a mass or nodule characteristic of IgG4-RD, 2) Serum IgG4 levels greater than 135 mg/dl, 3) Positivity for two of the following three criteria: (I) Dense lymphocyte and plasma cell infiltration with fibrosis; (II) Ratio of IgG4 positive plasma cells/IgG positive cells greater than 40% and the number of IgG4 positive plasma cells greater than 10 per high powered field; (III) Typical tissue fibrosis, particularly storiform fibrosis or obliterative phlebitis. In the diagnostic criteria, biopsy specimen is essential for making a definitive and probable diagnosis. IgG4 related pericarditis is difficult to diagnose histologically. We performed PET/CT for the determination of biopsy sites. IgG4-RD contains a variety of pathological changes which is dependent on the tissues involved, therefore this modality would be helpful. Horie, et al. reported cytological examination of the pericardial effusion.

Most patients with IgG4 related pericarditis show a rapid clinical response to steroid therapy and it possibly prevents the progressive involvement of other organs. However, if the steroid therapy delayed, the disease may lead to constrictive pericarditis. Constrictive pericarditis occurs in approximately 2%-3% of cases after pericarditis. Therefore, prompt therapy may suppress the progression to constrictive pericarditis.

Available treatments for inducing and maintaining remission are the same for all IgG4-RD phenotypes. Glucocorticoids (GCs) should be used as first line therapy to induce remission. In case of relapse, rituximab represents the most promising second line agent to re-induce remission. Maintenance of remission can be pursued with long term low dose GCs (oral prednisone 5-7.5 mg daily), Disease Modifying Anti-Rheumatic Drugs (DMARDs) or rituximab infused every six months.

In general, none of the available disease biomarkers alone can be considered a reliable mirror of disease activity. However, specific clinical scenarios in which these biomarkers might still be useful is growing numbers. There are traditional (elevation of serum IgG4 and IgE and peripheral blood eosinophilia, plasmablasts, PET/CT) and novel (autoantigens (prohibitin, annexin A11, laminin 511-E8 and galectin-3), soluble interleukin 2 receptor, memory B cells, activated Tfh2 cells, serum Enhanced Liver Fibrosis (ELF) score) markers and future studies are warranted which biomarker is suitable for monitoring in IgG4-RD activity.

CONCLUSION

The diagnosis of IgG4 related pericarditis is often difficult in terms of histologically analysis. PET/CT help identify IgG4 related organs and is useful to evaluate the efficacy of steroid therapy for IgG4 related disease. Most patients with IgG4 related pericarditis show a rapid clinical response to steroid therapy and it possibly prevents the progressive involvement of other organs. Prompt treatment contributes to improve the status of the disease.

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