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COMMENTARY

Alopecia and IgG4-related disease

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IgG4-related disease (IgG4-RD) is a recognized clinical affection characterized by the elevation of serum IgG4 levels and tissue infiltration of IgG4-positive plasma cells[1,2]. IgG4-RD causes inflammation with fibrosis and shows various symptoms corresponding to affected organs including pancreas, salivary grand, lacrimal grand, biliary tract, and peritoneum.

The comprehensive clinical diagnostic (CCD) criteria listed by Umehara[1]:

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1. Clinical study shows characteristic diffuse/localized swelling or masses in single or multiple organs

2. Hematological study shows elevated levels of serum IgG4 (135 mg/dL or higher)

3. Histopathological study shows the following two findings:

(i) Histological findings: marked lymphocyte and plasmacyte infiltration and fibrosis

(ii) IgG4-positive plasma cell infiltration: ratio of IgG4/IgG positive cell >40%, and IgG4-positive plasma cells/HPF >10

Of the above: When 1) + 2) + 3) are fulfilled, it is definite

When 1) +3) are fulfilled, it is probable

When 1)+ 2) are fulfilled, it is possible

However, it is important to differentiate from malignant tumors of each organ (cancer, lymphoma, *etc*.) and similar diseases (Sjogren's syndrome, primary sclerosing cholangitis, Castleman's disease, secondary retroperitoneal fibrosis, Wegener's granulomatosis, sarcoidosis, Churg–Strauss syndrome, *etc.*) with additional histopathological examination as much as possible

Even in the case that patients cannot be diagnosed with CCD criteria for IgG4RD, they may be diagnosed using organ-specific diagnostic criteria for IgG4RD.

By the way the skin disease is found in 4–7% of patients with IgG4-RD[3]. Dr. Tokura categorized IgG4-related skin disease based on the previous documentations and his experience, focusing on the aetiology and differential diagnoses[4]. He showed 7 types of skin lesions in IgG4-RD such as 1) cutaneous plasmacytosis, 2) pseudolymphoma and angiolymphoid hyperplasia with eosinophilia, 3) Mikulicz disease or IgG4-related dacryoadenitis and sialadenitis, 4) psoriasis-like eruption, 5) unspecified maculopapular or erythematous eruptions, 6) hypergammaglobulinaemia, purpura and urticarial vasculitis, 7) ischaemic digit.

Recently, alopecia is reported as one of skin manifestation of IgG4-

related disease (IgG4-RD)[3]. In the last issue of Trends in Immunotherapy, Dr. Yoshimasu reported the combination use of triamcinolone acetonide and immunotherapy as a new therapeutic option in alopecia totalis[5]. Although autoimmune mechanisms are considered as a major cause of alopecia, IgG4-related alopecia will suggest a new possibility of therapeutic approaches.

References

1. Umehara H, Okazaki K, Masaki Y, *et al*. Comprehensive diagnosis criteria for IgG4-related disease (IgG4-RD), 2011. Mod Rheumatol 2012; 22(1): 21–30. doi: 10.1007/s10165-011-0571-z.
2. Umehara H, Okazaki K, Nakamura T *et al*. Current approach to the diagnosis of IgG4-related disease – Combination of comprehensive diagnostic and organ-specific criteria. Mod Rheumatol 2017; 27(3): 381–391. doi: 10.1080/14397595.2017.1290911.
3. Ikeda T, Kaminaka C, Furukawa F. A case of alopecia as IgG4-related skin disease. Mod Rheumatol 2016; 23: 1–4. doi: 10.1080/14397595.2016.1233929.
4. Tokura Y, Yagi H, Yanaguchi H, *et al*. IgG4-related skin disease. Br J Dermatol 2014; 171(5): 959–967. doi: 10.1111/bjd.13296.
5. Yoshimasu T, Mikita N, Ikeda T, *et al*. Combination use of triamcinolone acetonide and immunotherapy as a new therapeutic option in alopecia totalis. Trends Immunother 2018; 2(1): 149. doi: 10.24294/ti.v2.i1.149.