Selective IgM Deficiency Accompanied with IgG4 Deficiency, Dermal Complications and a Bronchial Polyp

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Background: IgM deficiency is a rare primary immunodeficiency. As few studies of selective IgM deficiency have been reported among the various other types of primary immunodeficiencies, the detailed pathogenesis of this disorder remains to be elucidated.

Case Summary: We clinically analyzed a 37-year-old woman who presented with IgM and IgG4 deficiency and ectopic bronchial pneumonia, and investigated immunological functions. Occlusive pneumonia was repeatedly observed in the right S6 area, and bronchoscopy revealed a polyp in the right B6 orifice, which was later identified as a fibroepithelial polyp after transbronchial endoscopic polypectomy. Two months later, pneumonia involving the right inferior lobe developed. Systemic erythema and pigmentation with bleb formation were also observed on the skin, and were thought to be drug-induced exanthema following a biopsy. Serum levels of IgM and IgG4 were extremely low at 3.0 mg/dl and less than 2.0mg/dl, respectively. Circulating CD20 positive B cells were mildly reduced and memory B cells were markedly decreased. The majority of B cells expressed IgM on their surface. There were no abnormalities in cell counts of neutrophils, T cells, NK cells and monocytes. Chemotaxins, bactericidal activity and phagocytosis of neutrophils were normal.

Discussion: There have been no case reports of selective IgM deficiency with concurrent IgG4 deficiency, various dermal symptoms and a bronchial polyp, as demonstrated in our patient.

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