英文題目: Metachronous of IgG4 related disease: a case report
中文題目: 異時性IgG4相關疾病-個案報告
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Introduction

IgG4-related disease (IgG4-RD) is an immune-mediated disease which is gradually recognized by the world. IgG4-RD is commonly characterized by tumor-like lesion which infiltrate with lympho-plasmacytic cell, especially rich in IgG4 plasma cell. This disease course if often subacute, and be found incidentally through radiologic or unexpectedly in pathological specimens. IgG4-RD could involve single or multiple-organ.

Case Summary

A 55-year-old man who had history of aorta dissection initially presented with progressive painless jaundice and asthenia for 2 weeks. Abdominal CT scan revealed pancreatic head mass with biliary tree dilatation along with direct hyperbilirubinemia (Total bilirubin 23 mg/dl, Direct bilirubin 13 mg/dl) and elevated CA19-9 (7588.50 U/ml) ; He underwent Whipple procedure due to suspicion of pancreatic cancer, but pathology revealed only inflammatory cells infiltrated in the mass. Immunohistochemical study showed the plasma cells were positive for CD138 and IgG4. The ratio of IgG4-positive to IgG-positive plasma cells was beyond 60%, and more than 10 $IgG4_+$ plasma cells per high-power field; Serum IgG4 was 1800 mg/dl. The patient was discharged after operation 17 days later. Then, he took oral prednisolone (0.6 mg/kg/day) for 4 weeks and he tapered down without any medication control after 3 months.

After two years, he had bilateral submandibular swelling as well as elevated IgG and IgG4 level, (IgG: 8400mg/dl, IgG4: >4400 mg/dl), and decreased C3 and C4 level (C3:41.6mg/dl, C4:2.1mg/dl). Musculoskeletal ultrasound of bilateral submandibular gland showed multiple hypoechoic foci scattered against a heterogeneous background of salivary tissue. Anti-Ro, Anti-La, RF, and ANA were all negative. The patient was prescribed with oral prednisolone (0.6mg/kg/day) for one month without significant resolution of bilateral swollen glands. Therefore, the patient underwent steroid pulse therapy (methylprednisolone 500mg for three day). The swelling of involved organs subsided one week later, and the ultrasound also revealed the gland return to normal appearance with homogenous echotexture and

fine soft tissue echogenicity without any hypoechoic spots. After discharge, he refused maintenance steroid therapy due to afraid of side effects.

Three weeks after pulse therapy, he had cough, right upper chest wall pain and dyspnea on exertion. Chest radiograph showed patchy opacity at right upper lung field and diffuse alveolar infiltrates over bilateral lower lung fields. The clinical symptoms including chest wall pain and chest X ray did not improve after antibiotics. Bronchoscopic lavage revealed no pathogen or any intra-tracheal lesion. Finally, oral prednisolone (40mg/day) was prescribed, and the patient had dramatic improved in both radiography and symptoms.

Discussion

The case sequentially invaded pancreas, submandibular gland, and lung in different time. It was known glucocorticoids appear to be effective (0.6mg/kg/day initially, at least) in the majority of patients with IgG4-related disease, but disease flare are common. Maintains therapy (5mg/day) is necessary to reduce recurrence rate about 20-40%, but there was no known principle about the duration. In this case, sclerosing sialentitis and intertital pneumonia could be reversible, but consider his frequent recurrence situation and poor compliance to steroid, B-cell depletion with rituximab may appears to be a useful approach.

Conclusion

IgG-4 related disease could be synchronous or metachronous, and maintenance therapy including steroid or other immunosuppression agents are necessary. Furthermore, even though malignancy was highly suspect by tumor marker, image and clinical presentation, always keep in mind with differential diagnosis to avoid unnecessary surgery, especially without pathology confirmed.

Reference :

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