Three cases of IgG4-related sclerosing disease

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Introduction
IgG4-related sclerosing disease is a systemic disease associated with IgG4-positive lymphoid and/or plasmacytic infiltrations of many organs throughout the body. Infiltration of IgG4-positive plasma cells are often observed in salivary glands, thyroid, parotids, bile duct, and retroperitoneum. Scarring salivary glands (such as Mikulicz’s tumor), pancreatitis, pseudotumors (PT), primary sclerosing cholangitis (PSC), and retroperitoneal fibrosis may be IgG4-related diseases. We report three cases of IgG4-related sclerosing disease and its treatment.

Case 1: 56 years old, male

Present illness:
Bilateral submandibular swelling without pain, so he consulted an otolaryngologist. Therefore, in March 2008, he was introduced to us.

Past history:
nothing notable

Laboratory data:
IgM 125 IU/ml, IgA 183 IU/ml, IgG 1480 mg/dl, IgG1 660, IgG2 609, IgG3 7.5, IgG4 380 mg/dl.

Pathological finding:
Fibrous tissue with inflammatory tumor; treated with steroid.

Clinical course:
After swelling of salivary glands, IgG4 (+) plasmacytes were observed into the lacrimal glands, and sIL-2R 1690 U/ml.

Case 2: 47 years old, female

Past history:
hypertension

Laboratory data:
AMY 76 IU/L, ALP 136 IU/L, IgG 1480 mg/dl, IgG1 776 mg/dl, IgG2 702 mg/dl, IgG3 19.7 mg/dl, IgG4 531 mg/dl.

Clinical course:
Slight bilateral submandibular swelling. Bilateral lacrimal swelling. We will continue our observations, because we often see recurrences without steroid.

Case 3: 50 years old, female

Past history:
Dryness Thirst +/- Dry eye Dry mouth

Laboratory data:
CRP 7.5 mg/dl, IgM 50 mg/dl, IgA 60 mg/dl, IgG 400 mg/dl, IgG1 500 mg/dl, IgG2 300 mg/dl, IgG3 10 mg/dl, IgG4 70 mg/dl.

Clinical course:
After swelling of salivary glands, IgG4 (+) plasmacytes were observed into the lacrimal glands, and sIL-2R 1690 U/ml.

Discussion
IgG4-related sclerosing disease is a systemic disease associated with IgG4-positive lymphoid and/or plasmacytic infiltrations of many organs throughout the body. Cases of Mikulicz’s disease complicating IgG4-related diseases, such as AIP, are often reported. Although the complaint is swelling of the salivary glands, it often shows complications of IgG4-related systemic disease, such as AIP or PSC. Therefore, we must keep other organs in mind.

Treatment
Complications treatable for IgG4-related sclerosing disease is not well known, because only a few cases are reported. But whole body medical therapy was very effective, so we also have found.

In those cases, symptoms improved with steroid and deteriorated without steroid. We suggest that steroid is effective because not only salivary and lacrimal glands but also panocrine is improved.

Conclusions
In three cases of IgG4-related sclerosing disease with bilateral submandibular swelling, steroid therapy is very effective.

We must keep in mind other organs, because we often see complications with other systemic disease.