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A Case Report of Orbital IgG4 Related Disease

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***Abstract*—*IgG4 related disease is an uncommon systemic disorder which involves almost all organs in the body with characteristic fibrosis. The characteristic features are chronic non inflammatory lid swelling and proptosis. The term systemic condition IgG4-related disease, which is characterised by lymphocyte infiltration, plasma cell infiltration and consequent fibrosis in affected areas, is IgG4-related ophthalmic disease (IgG4-ROD). Systemic steroids are used as first line to decrease the lesion size, but relapse are more common after discontinuation*.

***Keywords*—** *IgG4, myositis, opthalmic disease, proptosis, periorbital swelling*.

I. INTRODUCTION

IgG4 related orbital disease will have characteristic orbital swelling and proptosis without signs of inflammation and elevation of serum IGg4 level with IGg4 infiltration in biopsy. It can occur in any age group. Male and female are equally involved. Any number of the orbital structures may be involved. The lacrimal glands, extraocular muscles, infraorbital nerve, supraorbital nerve, and eyelids are frequently affected structures. Additionally, it has been hypothesised that ligneous conjunctivitis may be a symptom of an IgG4-related illness (IgG4-RD). Systemic IgG4-related lesions might occasionally be present in these patients. There may also be IgG4-related abnormalities in the thyroid and pituitary. . IgG4-ROD often responds quickly to steroid therapy, as is the case with other symptoms of IgG4-related illness, unless considerable fibrosis has already taken place. Patients most commonly have associated allergic disease.

II. CASE PRESENTATION

A 28 year old male patient presented with complaints of bilateral orbital swelling, it developed gradually over a period of 7 months. No signs of inflammation, no visual disturbance.

Patient has no allergic history, no history of outside food intake. Routine blood investigations were normal.

On physical examination, both the eyes exhibited mild tenderness. He had 3.5mm proptosis in the left eye. Other physical examination findings were unremarkable. Complete blood count was normal.

Since the most common cause of orbital swelling is thyroid eye disease. But thyroid function test was normal. Since thyroid ophthalmopathy can rarely occur in euthyroid states, antiTPO antibody was done and it was negative.

MRI orbit showed increased ocular muscle thickness and Infra orbital nerve enlargement was present.

Serum albumin and immunoglobulin were evaluated. Serum IgG4 level was elevated-324mg/dl (normal: 4.8 - 105mg/dl). Biopsy of inferior recuts muscle revealed lymphoproliferative lesion with Mild fibrosis and IgG4+ plasma cells. Since it fits into the criteria of IgG4-RD, a diagnosis of IgG4 related ocular myositis was considered.

Patient was started on intravenous prednisolone 40mg for 1 week. Patient showed improvement as evidenced by decrease in orbital swelling. After 2 weeks steroids were tapered and was advised to continue maintenance dose of prednisolone 5mg/day. Patient's symptoms completely resolved after 6 months.

III. DISCUSSION

It is very much important to differentiate IGg4 related orbital myositis with idiopathic orbital inflammation, ocular marginal zone B cell lymphoma, and ANCA - related vasculitis. Histology is used to differentiate IgG4 related disease from other conditions. Mikulicz's disease is commonly associated with bilateral lacrimal gland lesion. Chronic periorbital edema and proptosis are the only significant feature of IgG4 related orbital myositis, without signs of inflammation. There may be mild restriction in ocular motility with no visual disturbance. Different degrees of lymphoplasmacytic infiltration with prominent sclerosing lesions or reactive lymphoid follicles are present in the histology of orbital IgG4-related illness. Additionally, eosinophilic infiltrations have been seen. Rarely, lymphoplasmacytic cells and macrophages harbouring eosinophilic material may infiltrate an orbital IgG4-related illness. Histologically, obliterative phlebitis is a hallmark of IgG4-related diseases in the body; however, it is uncommon in IgG4-related disorders of the orbit. IgG4-positive plasma cells are visible through immunohistochemical analysis which distinguishes IgG4-related disease from other inflammatory disorders originating from the ocular adnexa.

MRI is used to see the extent of inflammation. Infraorbital nerve enlargement (infraorbital nerve diameter is larger than the orbital nerve) is a sign for IgG4 related ophthalmic disease.

Systemic steroids, radiation, or rituximab are possible treatments for those with illnesses associated with orbital IgG4. It is challenging to evaluate the therapy outcomes by meta-analysis because studies on each of these treatments typically included only a small number of individuals. Once systemic steroid therapy, disorders caused by orbital IgG4 resolve, but recurrence is frequently seen after therapy is stopped. Relapses of lesions are seen in Mikulicz's illness after steroids are stopped. Prednisolone may therefore be best kept at 5 to 10 mg/day or combined with an immunosuppressant like azathioprine. Rituximab treatment prompts clinical and serologic improvement in patients with

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refractory IgG4-related disorders while relapse is also seen in some individuals with orbital IgG4-related disease after rituximab treatment is discontinued.

Systemic steroids are the main stay of treatment for IgG4 related disease. Relapse of symptoms is common after discontinuation of steroid. Other treatment includes radiotherapy or rituximab.

Recurrence is seen even after rituximab therapy. To prevent relapse prednisone can be combined along with any immunosuppressants.

IV. CONCLUSION

There are several unique features in orbital IgG4 related disease. IgG4 related disease should be considered in the differentials of lymphoplasmacytic or any sclerotic orbital lesions. Lacrimal gland and extraocular tissue are the commonly involved structures in IgG4-related diseases. It is important to start treatment immediately as it can lead to serious organ dysfunction but not every disease manifestation of IgG4 - related disease requires immediate treatment.

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