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A Rare Presentation of Igg4 Related Cavernous Sinus Disease– A Case Report

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1. INTRODUCTION

IgG4-related disease (IgG4-RD) is a chronic fibro-inflammatory condition characterized by mass-forming lesions in various tissues. The hallmark features of this disease include pathological lesions marked by dense lymphoplasmacytic infiltrates with numerous IgG4 plasma cells, typically accompanied by storiform fibrosis and a risk of organ damage[1].

IgG4-RD is an autoimmune disorder associated with elevated serum IgG4 levels and the infiltration of IgG4 plasma cells into affected tissues[2]. IgG4 is a subtype of immunoglobulin G (IgG), making up approximately 3-6% of total IgG[2]. Elevated serum IgG4 levels have been linked to several conditions, such as autoimmune pancreatitis, sclerosing cholangitis, Mikulicz disease, and retroperitoneal fibrosis, which are now collectively classified as IgG4-RD [2]. The organs most commonly involved include the pancreas, salivary glands, and biliary tract [2]. In the head and neck region, the salivary, lacrimal, and pituitary glands are frequently affected [2].

IgG4-RD affecting the cavernous sinus is quite rare. In this report, we present a case of a patient diagnosed with IgG4-RD involving the cavernous sinus after follow-up scan after 6 months duration. Imaging findings related to cavernous sinus involvement are limited to case reports, with very few cases previously documented.

2. METHODS

A 65 year old patient came with complaints of bilateral entropion with decreased visual acuity and complaints of oral numbness for 15 days (February 2024).The patient is known case of basal meningitis. No complaints of vomiting/seizures/weakness/deviation of angle of mouth, drooling of saliva. The patient was at first given differential diagnoses of Tolsa-hunt syndrome/Granulomatous infective/Non infective lesions since the MRI Brain plain and contrast study done on February 2024 showed enhancement in the region of left cavernous sinus with extension till the superior orbital fissure and into the foramen rotundum and ovale . Then the patient was treated with steroids and the symptoms resolved. These complaints reoccurred in July 2024 then on further radiological and laboratory evaluation a conclusive diagnosis was made and patient was treated accordingly.

MRI Brain plain and contrast study was repeated in July 2024 and showed bilateral involvement of the cavernous sinus.

Laboratory investigations revealed Serum IgG4 levels to be raised > 135 mg/dl has 99% specificity for IgG4-RD.So conclusive diagnosis of IgG4 related disease was made. Elevated total IgG and IgE, peripheral eosinophilia, ESR (Erythrocyte Sedimentation Rate), and CRP (C -reactive protein) are elevated but are nonspecific findings.

3. RESULTS

The patient is currently undergoing steroid therapy with drastic improvements in the clinical symptoms with decrease in the Serum IgG4 levels.

4. DISCUSSION

IgG4-related disease is an autoimmune condition characterized by the infiltration of IgG4 plasma cells into various tissues, leading to fibrosis[2].IgG4-related disease (IgG4-RD) is a multi-organ condition primarily affecting the pancreatic and hepatobiliary systems[4]. Isolated involvement of the head, neck, and brain is relatively rare. The first reported case of IgG4-RD affecting the lateral skull base appeared in 2012, while isolated invasion of the cavernous sinus is even less common[4]. Studies indicate that the pancreas is generally the most affected organ, but IgG4-RD can potentially involve nearly every organ in the body, including the skull base[4]. This condition predominantly occurs between the sixth and seventh decades of life and is associated with lymphadenopathy, typically responding well to steroid therapy[4].

Radiologically, IgG4-related mass lesions, like in our case, typically appear as homogeneously enhanced lesions that are isointense to hypointense on T1-weighted images and hyperintense on T2-weighted and FLAIR images[4]. In our patient, MRI scans post-remission showed T2-weighted hyperintensity in the bilateral cavernous sinus and superior orbital fissure, whereas the previous scan showed involvement only in the left cavernous sinus[4]. Following treatment with prednisone at 40 mg/day for 2–3 weeks, tapering to 5 mg/day over 3–6 months was implemented[4].

It is crucial for patients with IgG4-RD to receive treatment, as untreated cases can lead to extensive organ fibrosis[4]. Surgical intervention may be necessary to prevent recurrent episodes, but it should be considered only for patients who are suitable candidates and when steroid therapy is ineffective[4].

In addition to corticosteroid therapy and surgery, various other treatment regimens are currently in practice, with some still undergoing trials[3]. One option includes B-cell depletion therapy, administered at 1 g intravenously (IV) every 15 days for a total of two doses[3]. Patients receiving B-cell depletion often experience a rapid clinical response and a decrease in serum IgG4 levels; however, the reduction in tissue IgG4 and plasma cells is not significant[3]. Bortezomib, a proteasome inhibitor known for its cytotoxic effects on plasma cells and approved for multiple myeloma, has also shown effectiveness in treating IgG4-related lung and orbital disease, according to reports[3].

Comprehensive clinical diagnostic criteria for IgG4-RD[1]

1.Clinical examination showing characteristic diffuse or localized swelling or masses in single or multiple organ[1].

2.Elevated serum IgG4 concentrations >135 mg[1].

3.Histopathologic examination showing[1].

a. Marked lymphocyte and plasmacyte infiltration and fibrosis

b. Infiltration of IgG4 + plasma cells: ratio of IgG4 +/IgG + cells >40% and

> 10 IgG4 + plasma cell

5. SUMMARY

This case underscores the importance of imaging findings in association with clinical and lab parameters in IgG4-RD patients.

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Declaration of patient consent:

The authors declare that informed consent was obtained from the patient or their legal representative for the publication of this case report. The patient has been informed about the nature of the report, its purpose, and the potential for identification through the details provided. All personal identifiers have been removed to ensure confidentiality.

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