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A Rare Presentation of IgG4-Related Disease as a Nasopharyngeal and Oropharyngeal Fibrosing Mass – A Case Report

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INTRODUCTION

- Immunoglobulin G4-related disease (IgG4-RD) is a fibroinflammatory condition characterized by tissue infiltration with lymphocytes and IgG4-secreting plasma cells.
- The clinical presentation of IgG4-RD is highly heterogeneous, often mimicking malignancies, infections, or other autoimmune diseases, making diagnosis challenging.

CASE DESCRIPTION

- We report a rare case of IgG4-RD presenting as a nasopharyngeal and oropharyngeal mass in a 70-year-old Malay woman.
- She presented with a 3-month history of painful neck swelling and weight loss, without other connective tissue disease (CTD) symptoms.
- A neck CT scan revealed an enhancing soft tissue mass (2.9 × 2.5 × 4.7 cm) in the left nasopharyngeal and oropharyngeal regions, suspicious for carcinoma with possible nodal metastasis.
- Initial transoral retropharyngeal biopsy showed fibromuscular tissue with neurovascular bundles and mild to moderate inflammatory infiltrates.
- Immunohistochemistry revealed a mixed population of B and T cells without evidence of malignancy or lymphoma.
- CTD workups were positive for ANA (titer 1:320) but negative for dsDNA and ENA panel, with normal C3/C4 levels. Serum total IgG was elevated at 1800 mg/dL.
- Infectious causes including tuberculosis were ruled out.
- Due to diagnostic uncertainty, a second biopsy was performed with special IgG4 staining.
- Histology demonstrated a reactive lymph node and inflamed, fibrotic fibroadipose tissue infiltrated by IgG4-positive plasma cells (CD138+), with an IgG4/IgG ratio of 50%, confirming the diagnosis of IgG4-RD.
- The patient was treated with oral prednisolone (0.5 mg/kg) and azathioprine (2 mg/kg).
- Follow-up MRI of the neck after six months showed significant reduction of the mass and resolution of the enlarged cervical lymph nodes.

DISCUSSION

Diagnosis of IgG4-RD in this patient was supported by:

1. Histopathological Evidence: Biopsy confirmed fibroadipose tissue with dense lymphoplasmacytic infiltration and fibrosis. Immunostaining revealed IgG4-positive plasma cells (CD138+) with an IgG4/IgG ratio of 50%, a threshold strongly indicative of IgG4-RD.
2. Total IgG was elevated at 1800 mg/dL.
3. Exclusion of Other Causes: Negative workups for malignancy, tuberculosis, and connective tissue diseases.
4. Dramatic response to steroids: Reduction of inflammatory markers (ESR and CRP) and size of the fibrosing mass after treatment with steroids.

Management:

- Treatment of IgG4-related disease primarily involves glucocorticoids as first-line therapy, often inducing rapid remission.
- Steroid-sparing agents like azathioprine, mycophenolate mofetil, or methotrexate are used for maintenance or in relapsing cases[1].
- Rituximab, an anti-CD20 monoclonal antibody, is effective in refractory or relapsing disease[2].
- Early therapy prevents irreversible fibrosis and organ damage.

CONCLUSION

- Histopathological evaluation with IgG4 immunostaining is crucial in establishing the diagnosisof IgG4-RD.
- Elevated serum IgG4 levels and a favorable response to corticosteroids support the diagnosis.
- Early recognition of IgG4-RD is vital, as timely treatment with corticosteroids and immunosuppression like azathioprine can lead to dramatic clinical improvement and avoid unnecessary interventions.

References:

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