IgA Nephropathy and Nasal Type Extranodal NK/T Cell Lymphoma: A Case Report and Review of Literature

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Case Description

History

- A 49 y/o Hispanic female with no significant PMHx presents with
  - progressively worsening headaches for 2 months,
  - low grade fever,
  - foul smelling nasal discharge,
  - left periorbital swelling and left eye tearing.
- Treated for bacterial sinusitis ~1 month prior, no relief.
- Negative ROS, no allergies, no pertinent family hx
Physical Exam

- **Vital Signs**: BP 104/62 mmHg, PR 105/min, T 99 °F, RR 20/min, SpO2 97% on room air
- **HEENT**: EOMI, mild periorbital swelling, blood-tinged purulent drainage from nares, tender sinuses bilaterally
- **Cardiac**: Tachycardic, normal S1 & S2
- **Rest of the exam otherwise unremarkable**
Laboratory Data

- Hgb 8.5 mg/dL
- CRP 7.0 mg/dL
- ESR 67 mm/hr
- Urinalysis: Microscopic hematuria
- CMP normal, c-ANCA and other serologies negative
Imaging
Fig. Selected axial images from CT of facial bones with contrast, showing soft tissue mass in right nasal cavity and extending to left orbit
Clinical course

- Bilateral nasal endoscopy and biopsy
  - Advanced necrosis of intranasal structures
  - Biopsy extensive bone and soft tissue necrosis
- Patient started on treatment for Wegener's granulomatosis
  though serology was negative
- Renal biopsy showed IgA Nephropathy
Clinical course

- No clinical improvement after 14 days.
  - Patient had repeat, broader tissue biopsies.
  - Pathology this time revealed ENKTL, Nasal type.
- Patient was started on chemo-radiation.
- Unfortunately, patient passed away 6 weeks later from disseminated disease.
IgA Nephropathy and Nasal type Extranodal NK/T Cell Lymphoma (ENKTL): A review of literature
Introduction

IgA nephropathy (IgAN)

- Most common cause of primary glomerulonephritis worldwide.
- Men more than women, all age groups
- Can be primary (idiopathic) or secondary (cirrhosis, IBD, malignancy)
- Rarely associated with lymphomas, with only 9 cases reported (3 Hodgkin’s, 4 Mycosis fungoides, 1 MALT-L, 1 ENKTL)
Nasal Type ENKTL

- An aggressive type of NHL involving nasal/paranasal structures
- Characterized by an extensive vascular destruction and tissue necrosis
- Rare in the US, commonly seen in Latin America and Asia
- Invariably associated with EBV infection
Pathogenesis

- Overproduction, defective clearance, altered mucosal immunity implicated in IgAN.
- Exact pathogenesis when associated with lymphomas is unclear.
- Mucosal necrosis and ulceration $\rightarrow$ hyperactivity of local mucosal immune system $\rightarrow$ IgA overproduction $\rightarrow$ IgAN.
- Per Talpur et al, 24.8% of 254 patients with cutaneous T-cell lymphoma had elevated IgA levels, suggesting a role for overproduction of IgA.
Clinical Presentation

- IgAN is often clinically silent.

- 9 reported cases of IgAN associated with lymphomas
  - All had hematuria
  - 6 had proteinuria
  - 6 had renal failure
  - Infiltration by lymphoma generally uncommon

- The patient with IgAN and nasal type ENKTL presented with hematuria, proteinuria and renal insufficiency.
Renal biopsy needed for definitive histologic diagnosis.

Fig. A glomerular tuft with mild global mesangial hypercellularity (H&E, x400)

Fig. Immunofluorescence staining of a single glomerulus, in which actual IgA deposits have been stained fluorescent green (x400)
Treatment

- Goal is to relieve symptoms, slow progression and prevent complications of IgAN.

- Roles of immunosuppressive agents, ACE inhibitors or ARBs, fish oil, and Vitamin E have been described.

- Treatment of the primary disease process may benefit in improvement, or even reversal, of secondary IgAN.
Treatment

- Complete renal recovery achieved in 4 of 5 lymphoma patients following treatment with chemotherapy, with or without radiotherapy or steroid treatment.

- In the first case report of IgAN and nasal type ENKTL, serum creatinine normalized after chemotherapy; however, hematuria and proteinuria persisted.

- In our patient, microscopic hematuria resolved after starting chemotherapy.
Conclusion

- To the best of our knowledge, this is the second case report illustrating association between IgAN and nasal type ENKTL.

- The improvement in hematuria following chemotherapy likely supports a probable pathogenetic relationship.

- The spectrum of lymphomas associated with IgAN should be expanded to include nasal type ENKTL, and physicians should be aware of this association.
Reference


Thank you!