Extranodal Natural Killer/T-Cell Lymphoma Presenting as a Palatal Ulcer

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History of Present Illness

• 32-year-old caucasian male referred to our outpatient otolaryngology clinic for painful mouth ulcer

• First discovered small palate ulcer 1 month prior

• Began with “tickle” in his throat and within days became severe odynophagia, dysphagia, otalgia, and fatigue

• He also noticed a ruborous rash that began on his arms and spread to his abdomen, chest, back, buttocks, and legs
History of Present Illness

• Primary care physician prescribed him Augmentin, with no improvement noted

• 2 weeks after symptoms began, presented to Emergency Department (ED)

• CT maxillofacial with contrast and Complete Blood Count (CBC) performed in ED
  • 2.5 cm retention cyst right maxillary sinus
  • CBC unremarkable

• Referred to ENT
Past Medical/Surgical History

• Attention deficit hyperactivity disorder
• Anxiety disorder
• No surgical history
Social/Family History

- Chiropractor
- Married
- Denies alcohol, tobacco, or illicit drug use
- History of testicular cancer in father
Medications/Allergies

- Zyrtec (cetirizine)
- Cepacol (benzocaine)
- Ibuprofen
- NKDA
Physical Exam

- General: Well-appearing male in no acute distress
- Vitals within normal limits
- **HEENT:** fibrinous, erythematous, ulcerative oral lesion, midline involving the hard and soft palate
- Neuro: CNs II-XII intact, alert and oriented to person, place, time
- Extremities/Trunk: patchy, circular, non-palpable, ruborous, maculopapular rash over the trunk and upper and lower extremities
Palate Lesion (initial presentation)
Initial Assessment/Plan

- Likely autoimmune process, cannot rule out malignancy
  - Sarcoidosis
  - Granulomatosis with polyangiitis (formerly Wegener's granulomatosis)
  - Psoriasis
  - Pemphigus vulgaris
- Palate biopsy scheduled one week out
- Placed on prednisone taper and Magic Mouthwash
- Referral to dermatology for rash
One Week Follow Up

- Patient’s rash improving with steroid taper
- Oral symptoms slightly improved
- Palatal biopsy performed
  - 3 mm punch biopsy
  - Pathology revealed lichenoid lymphocytic infiltrate suggesting the possibility of lichen planus and lichenoid medication reaction
    - No evidence of dysplasia or malignancy
- CBC, LFTs, ANA, c-ANCA, p-ANCA, ACE all negative
- 3-4 week follow up scheduled
Four Week Follow Up

- Symptoms worsened
  - Horrible odor coming from mouth
  - Significant oral pain limiting ability to eat
  - Liquid coming through nose while drinking
- Physical exam revealed extension of ulcer and bony erosion measuring 5.5 x 3.5 cm
Palate Lesion (2 months after symptoms began)
Timeline

4 weeks
Four Week Follow Up Cont.

- Repeat palatal biopsy and left septal biopsy performed
  - 4 mm punch biopsy
  - Pathology from palate and septum revealed extranodal Natural Killer/T-Cell Lymphoma
  - Immunohistochemistry
    - **Strongly EBV positive**, CD3 and CD56 positive
  - Immediate hematology/oncology referral made
Hematology/Oncology

- Workup included Epstein-Barr virus by PCR, C reactive protein, HIV, LDH, CBC, and CMP.
  - All negative/within normal limits except WBC 3.9

- PET/CT scan and MRI of the head for complete radiographic staging workup and a baseline bone marrow biopsy were done

- No evidence of bone marrow involvement
PET/CT Scan

- Highly FDG avid:
  - **Invasive nasopharyngeal soft mass**, max SUV 21.5
  - **Bilateral cervical lymphadenopathy**, left level 2 cervical node measuring 1.8 x 1.4 cm with max SUV of 13.0 and right level 2 cervical node measuring 1.5 x 1.5 cm with max SUV of 7.0
  - **Multiple osseous lesions** highly suspicious for neoplastic involvement
    - Right clavicular head max SUV 10.2, Right distal clavicle max SUV 10.5, Sternal body max SUV 9.8, right patella max SUV 15.9, right distal fibula max SUV 12.0
    - Left patella max SUV 17.5, and left distal tibia with max SUV 6.6
    - Patchy ground-glass opacities within the right lower lobe
  - **Stage IV** disease given metastases
Treatment

- Chemotherapy was the modified SMILE (mSMILE) regimen; **Palliative**
  
  - *(Steroid=dexamethasone 40 mg IVPB days 2-4, methotrexate 2000 mg/m² day 1, ifosfamide 1500 mg/m² days 2-4, pegylated-L-asparaginase 2500 IU/m² IVPB day 8, and etoposide 100 mg/m² days 2-4)*

- The patient underwent **3 cycles of mSMILE** along with radiotherapy to bulky original sites

- Subsequent PET/CT scan revealed a dramatic response with **no further FDG avid disease**. Although high dose chemotherapy and stem cell transplantation as consolidation was recommended, the patient opted for close interval surveillance instead
Palate Lesion (after 3 rounds of chemotherapy)
Treatment

- Patient underwent palatal reconstruction at an outside facility
- Additionally, he was fitted for dental retainer after the defect had healed from surgery
- Unfortunately, the patient was lost to follow up and his current status is unknown
Post-Op Palatal Recon. and Dental Retainer
Discussion

- Extranodal natural killer/T-cell lymphoma (ENKTL), nasal type, is a rare form of non-Hodgkin lymphoma characterized by local necrosis and angioinvasion

  - Also termed “lethal midline granuloma”

- Most cases are Epstein-Barr virus (EBV) positive and possess the cell markers of true natural killer cells: CD2, cytoplasmic CD3, and CD56

- Outside of Asia and Central/South America it is very rare

- Median age of onset is in the sixth decade of life with a male-to-female ratio of 2:1
Discussion

• Clinically, it usually presents as nasal obstruction, epistaxis, mass, or septal perforation

• Diagnosed with biopsy and immunohistochemistry (EBV, CD2, CD3, and CD56)

• Treatment is chemoradiation

• Prognosis is poor, median survival time is reported as 12.5 months

• In the English literature, prior to this, only two cases have been reported with palatal ulceration without accompanying nasal symptoms

• Case highlights importance of keeping malignancy high in the differential of a patient with a rapidly destructive palatal lesion
Timeline

4 weeks
Timeline

mSMILE
3 rounds
Timeline

Palatal reconstruction

Dental retainer
References


