Nasal-type NK/T-cell Lymphoma in a Patient with Hidradenitis Suppurativa: A Case Report

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Introduction
Nasal-type NK/T-cell lymphoma (NKTL) is a rare disease characterized by the rapid necrosis of the nasal cavity and adjacent structures. While early diagnosis is critical, such expediency is rare due to substantial variation in patient presentation and the prominence of nonspecific nasal symptoms. Frequent misdiagnosis of this condition leads to delayed treatment and high mortality rates.

Purpose
To describe an emblematic case of nasal-type NKTL in a patient whose disease was initially attributed to his pre-diagnosed hidradenitis suppurativa (HS).

Case Overview
A 38-year-old male presented with several weeks of progressive nasal swelling and redness, which he initially attributed to his pre-existing HS and self-treated with antibiotics he already had on hand. The patient was later prescribed several courses of increasingly broad-spectrum antibiotics without improvement in his symptoms. Alternative non-infectious diagnoses were only given special consideration once the tumor had grown to involve nearby structures including the maxilla, nasal sinuses, and oropharynx with characteristic erosion of the hard palate.

A Note on Hidradenitis Suppurativa
Hidradenitis suppurativa (HS) is an inflammatory disease of the follicular epithelium. It presents with recurrent skin lesions: superficial abscesses, sinus tracts, and erythematous papules and plaques. HS is seen most often in the axillary, inguinal, and inframmary regions, but can also occur on the back, waistline, and rarely, the face (Figure 1). It is treated with oral antibiotics.1

Case Presentation and Timeline
A 38-year-old male with a history of hidradenitis suppurativa (HS) is admitted from the emergency department with a two-month history of progressive nasal swelling and redness.

1st Presentation
• The patient had first presented to his PCP after 3 weeks of worsening edema, erythema, and skin irritation
• He initially attributed his symptoms to pre-existing hidradenitis suppurativa (HS) and self-treated with antibiotics (prescribed for his HS)
• His PCP prescribed cephalexin for presumptive facial cellulitis, with minimal relief

2nd Presentation
• Worsening edema soon led to complete nasal obstruction prompting the patient to present to local urgent care
• A CT was obtained, which showed a mass filling the nose and nasopharynx (Figure 2)
• Given growing concern for an atypical infection and possible septal abscess, the patient’s antibiotic coverage was broadened to ceftriaxone and TMP/SMX

3rd Presentation
• Several weeks later, the patient presented to his local emergency department with worsening pain and redness of his hard palate as well as dysphagia, malaise, and recurrent fevers
• Past medical history was significant for HS of the bilateral axilla; he also endorsed a remote history of drug use
• Exam findings: vital signs stable; indurated scalp mass involving the entire nose, sessile ulcerative changes of the nasal turbinates (Figure 3)
• Labs unremarkable, including normal WBC and CRP
• MRI imaging was obtained (Figure 4), which demonstrated an infiltrative mass involving the nose, sinuses, oropharynx, and soft and hard palate, and maxilla—findings concerning for a sinonasal lymphoma
• After a failed attempt at a bedside intraoral biopsy, the patient was transferred to a tertiary care facility

The patient was taken promptly to the OR for an operative biopsy which revealed a high-grade lymphoblastic lymphoma, suspicious for nasal-type NK/T-cell lymphoma. He was transferred to medical oncology for chemoradiation therapy. Less than three months after diagnosis, his symptoms had resolved.

Discussion
Nasal-type NKTL is a rare type of non-Hodgkin lymphoma. Between 2005 and 2014, only 718 cases were diagnosed in the US according to the SEER database.2 Poor recognition of this disease has led to frequent delays in diagnosis and high mortality rates, with an estimated 5-year survival of 10-40%.3

There are striking geographic differences in incidence of this malignancy. In South America and Asia, nasal-type NKTL accounts for 3-10% of all non-Hodgkin lymphomas whereas this figure is reduced to less than 1% in Europe and the U.S.4 Nasal-type NKTL is seen more in males than females, particularly those between 40 to 50 years old. The pathogenesis of this disease is unknown, though studies have found an association between NKTL and the p53 and c-KIT gene mutations. There is also an association with Epstein-Barr virus (EBV) infection.5

Based on the typical presenting symptoms of nasal edema, swelling, obstructing, and episodic, this disease is frequently misdiagnosed as chronic sinusitis or a bacterial infection. However, systemic disease findings (eg, fever, weight loss, etc.) are typically absent in NKTL patients.6 Other comorbidities (in this patient’s case, hidradenitis suppurativa) can also misdirect diagnosis.

Workup of suspected nasal-type NKTL should include prompt endoscopy with biopsy. Histology will reveal coagulative necrosis and vascular invasion by tumor cells. CT or MRI should be performed to determine tumor extension. Current treatment recommendations favor L-asparaginase-based chemotherapy followed by radiation. Reconstructive surgery may be required.1,4

Figure 1. Hidradenitis suppurativa of the axilla (left) and perauricular area (right)6

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References