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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 strong 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 mostly in the axillary, inguinal and inframammary regions, but can also occur on the back, waistline, and rarely, the face (Figure 1). It is treated with oral antibiotics.¹

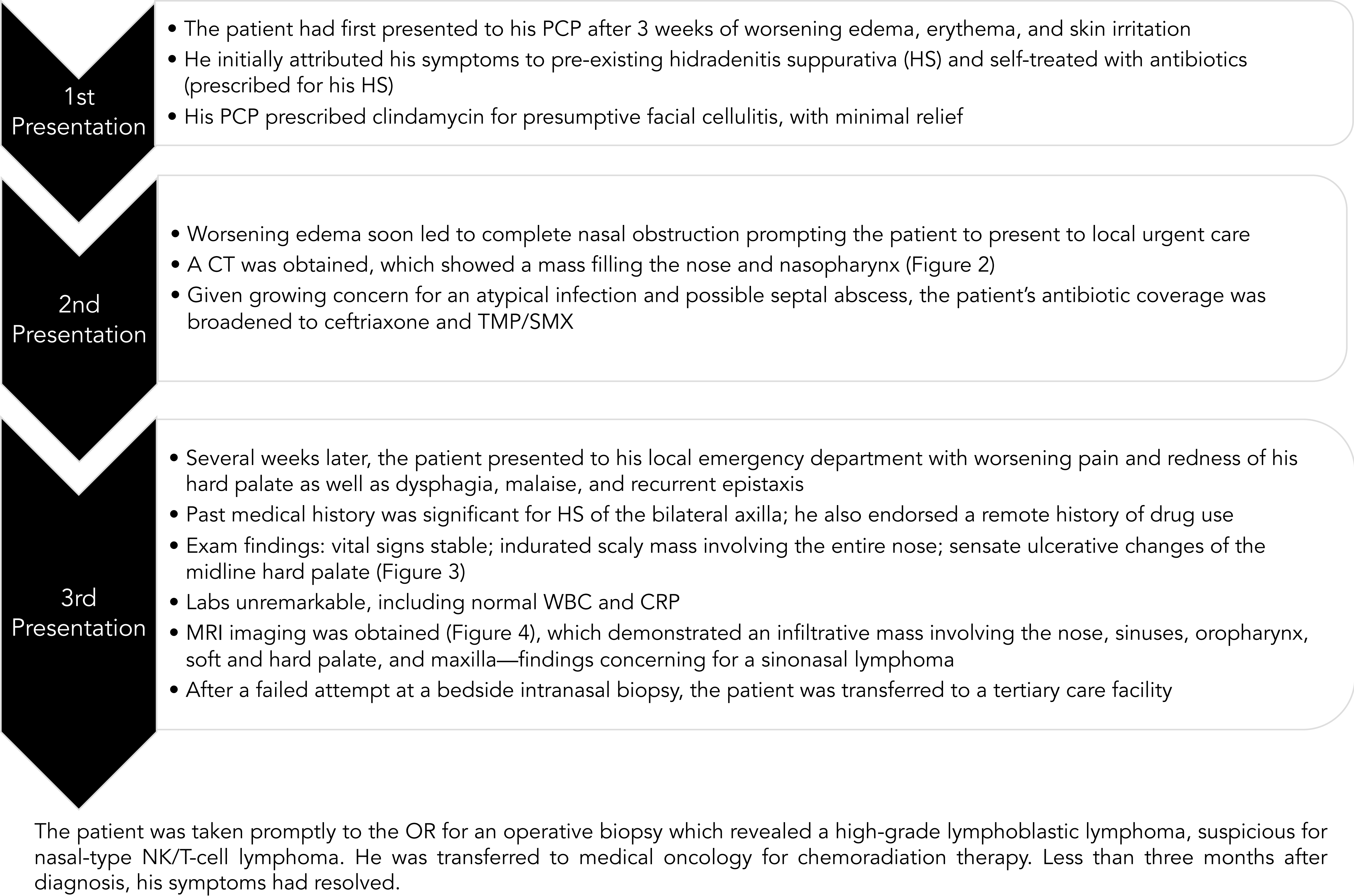


Figure 1. Hidradenitis suppurativa of the axilla (left) and periauricular area (right)⁵

Disclosures: The authors have no conflicts of interest to disclose.

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.



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.¹ Poor recognition of this disease has led to frequent delays in diagnosis and high mortality rates, with an estimated 5-year survival of 10-45%.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 erythema, swelling, obstruction, and epistaxis, 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.⁵ 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.^{3, 6}

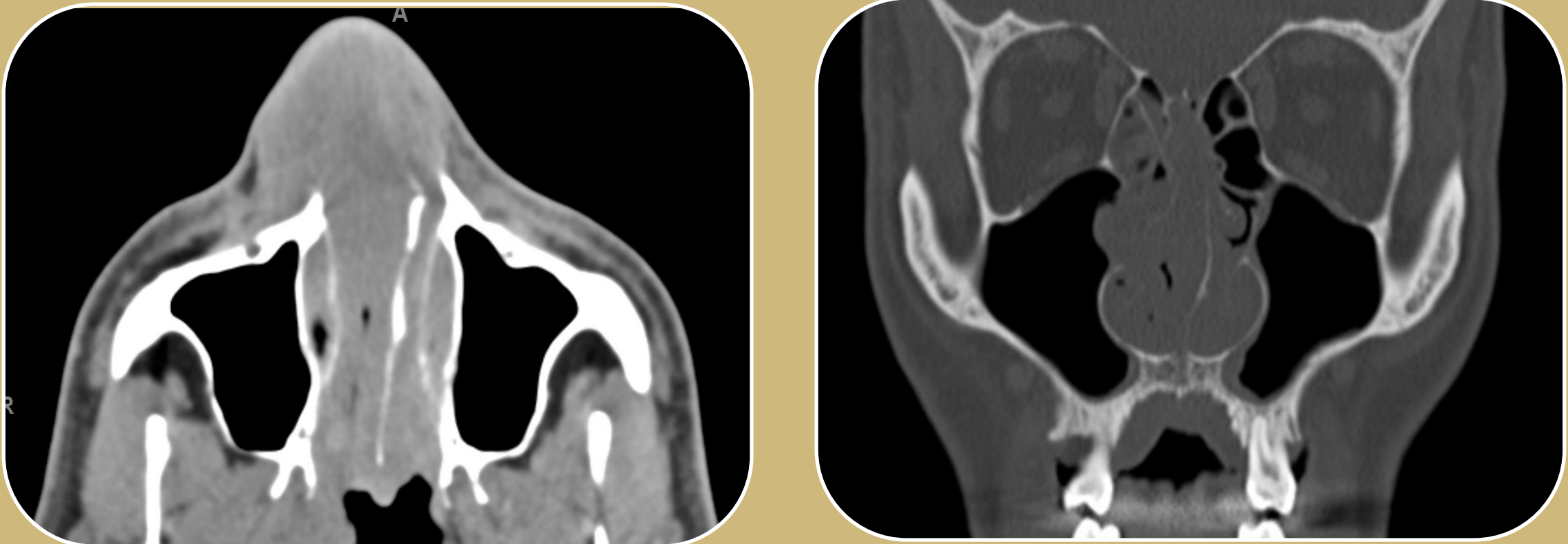


Figure 2. Axial soft tissue window (left) and coronal bone window (right) CT face with contrast

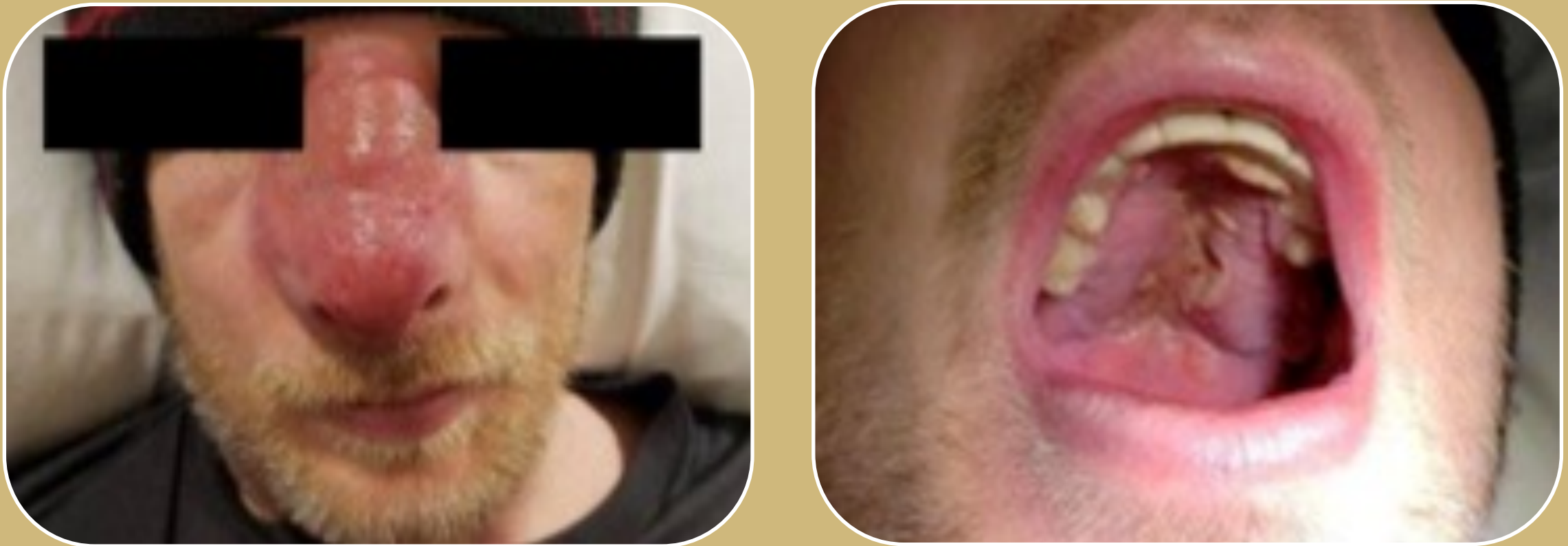


Figure 3. Pre-treatment facial (left) and intraoral (right) exam

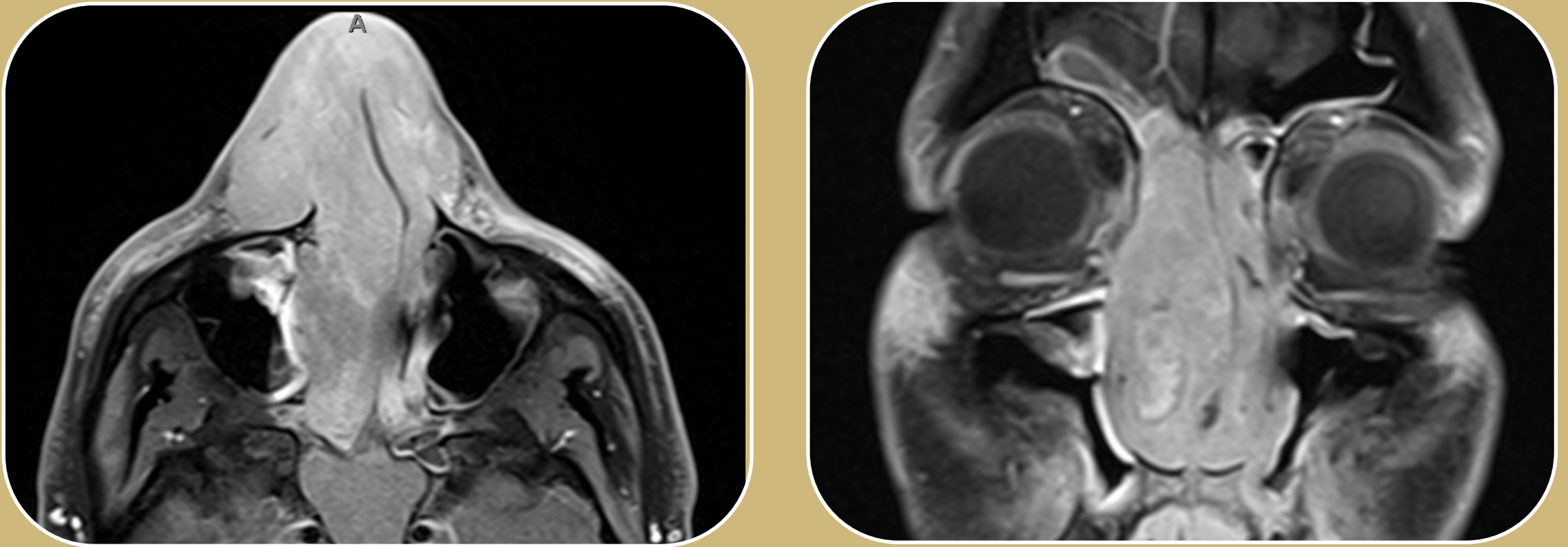


Figure 4. Axial (left) and coronal (right) T1 MRI with gadolinium obtained 1 month after initial CT

Conclusion

Nasal-type NK/T-cell lymphoma is a rare and often misdiagnosed disease. In this case, the patient initially attributed his symptoms to a dermatological condition and was repeatedly misdiagnosed with facial cellulitis. Nasal-type NKTL was suspected only when the patient developed necrosis of the hard palate. This case emphasizes the need for a broad differential when evaluating nasal inflammation without systemic signs of infection, particularly when symptoms progress despite antibiotic therapy.

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