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CASE REPORT

Extra nodal NK/T cell Lymphoma, Nasal type: Atypical Presentation of Rhinosinusitis cannot be Missed!

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Abstract

Extra nodal NK/T cell lymphoma (ENKTCL) nasal type is an Epstein Barr virus (EBV)-associated lymphoma of NK cells or T cells, predominantly involving extra nodal tissue. It most commonly affects the upper aerodigestive tract, including the nasal cavity, nasopharynx, paranasal sinuses, and palate. We report a case of a 50-year-old man with a blocked right nasal passage associated with unilateral right facial pain and swelling extending down to the neck. Endoscopic and radiological examinations revealed the presence of a soft tissue lesion within the right nasal cavity. Histopathological examination confirmed the diagnosis of ENKTCL nasal type.

Keywords: *Epstein Barr Virus; Extra nodal NK/T cell lymphoma nasal type; Rhinosinusitis.*

Introduction

Malignant lymphomas constitute around 4% of cancers in adults and are divided into Hodgkin lymphoma (HL) and non-Hodgkin lymphoma (NHL). About 90% of lymphomas are NHLs [1]. In adults, 85% of lymphomas are B-cell lymphomas, while only 15% are T-cell lymphomas. Extra nodal NK/T cell lymphomas (ENKTCL) constitute approximately 10% of T-cell lymphomas, making them relatively rare. They were previously referred to as 'lethal midline granuloma' due to their involvement in the midline facial area. East Asians and Native Americans in Central and South America are the most affected populations [2].

Case report

A 50-year-old fisherman, a chronic smoker with no known medical illnesses or drug allergies, presented to the ear, nose, and throat (ENT) clinic with a right nasal blockage persisting for the past month. Initially, he experienced intermittent fever for a week, accompanied by persistent rhinorrhea characterized by thick yellowish discharge, before progressing to right nasal blockage. A few days later, he began to develop right facial pain and swelling extending down to the neck. Apart from these symptoms, he denied experiencing epistaxis, frequent nasal itchiness or sneezing, cough, loss of weight, loss of appetite, night sweats, dysphagia, odynophagia, or shortness of breath. He had no history of previous contact with persons infected with tuberculosis. Despite receiving symptomatic treatment from two different local general practitioners, which included completing two courses of antibiotics, his symptoms continued to worsen.

On examination, there was diffuse unilateral right facial swelling. It was tender on palpation, firm, non-erythematous, extending down to the level I and II neck regions with a few palpable lymph nodes. Otherwise, the facial nerve was intact. In terms of the oropharynx, there was no trismus, the uvula was centrally located. However, a bulge was seen over the right soft palate. Other systemic examinations were unremarkable. All blood investigations showed normal parameters, including a full blood picture. Contrast-enhanced computed tomography (CECT) scan of the head and neck revealed features that may represent sinonasal malignancy. This was causing severe airway narrowing with nodal metastases and diffuse skin inflammatory changes [Figure 2(a) and 2(b)].

Subsequently, the patient underwent nasopharyngoscopy examination, revealing a mass in his right nasal cavity [Figure 1]. A biopsy of the right nasal cavity mass was performed. Histopathologic findings revealed predominantly necrotic tissue with a few atypical lymphoid cells present [Figure 3(a) and 3(b)]. Immunohistochemical studies confirmed the presence of these atypical cells, which were positive for CD56, CD3, and TIA. The Ki67 proliferative index was estimated to be approximately 50%-60%, and no granuloma was seen. Epstein-Barr encoding region (EBER) testing was positive by in situ hybridization (ISH) [Figure 4 (a-e)]. Consequently, the final diagnosis of extra nodal NK/T-cell lymphoma nasal-type was established. Unfortunately, despite receiving chemotherapy, the patient succumbed to multiple organ failure one month following his initial visit.

Discussion

Sinonasal NK/T-cell lymphoma is known to be one of the most aggressive lymphomas, causing progressive midline destruction of the nasal or mid-facial bones [3]. Although generally rare, sinonasal NK/T-cell lymphoma accounts for 2.6% – 6.7% of all lymphomas, but less than 0.5% of lymphoma cases in Western populations [4]. In Asians, after the gastrointestinal tract, the sinonasal tract is the second most common site of extranodal lymphoma [5,6]. Common presenting symptoms include nasal obstruction, rhinorrhoea, epistaxis, and, rarely, facial swelling and orbital symptoms such as ptosis or limitation of eye movement [5]. These symptoms and signs may

overlap in patients with rhinosinusitis, as they can present with similar symptoms of nasal blockage or obstruction with nasal discharge, either purulent or greenish, facial pain, smell disturbances, and fever. This patient experienced worsening and persistent symptoms for more than 10 days but less than 12 weeks, fitting the criteria for acute rhinosinusitis. At the same time, he presented with right-sided unilateral facial swelling with facial pain, together with neck swelling after having persistent unilateral right nasal blockage, which is not a typical presentation of rhinosinusitis. This notable facial pain and swelling, which extends down to the neck, should prompt consideration of additional differential diagnoses beyond acute rhinosinusitis alone.

This case report highlights two important points. Firstly, sinonasal carcinoma, which in this case is ENKTCL of nasal type, should be considered in patients presenting with atypical rhinosinusitis symptoms. Secondly, early referral should be done as soon as possible if the symptoms persist despite adequate and optimal therapy to rule out other important differential diagnoses.

This illustrates the challenges involved in making a diagnosis of ENKTCL nasal type, as its signs and symptoms often mimic those of rhinosinusitis [6]. Sands et al observed in a series of four patients that extra nodal nasal NK/T-cell lymphoma being mistaken for chronic sinusitis resulting in therapeutic delays in these patients (mean time to diagnosis = 21.5 months) [7]. In our case, this patient was initially treated for rhinosinusitis, leading to delay in establishing the diagnosis of ENKTCL nasal type. Generally, the prognosis for extra nodal NK/T cell lymphoma is poor despite localized disease at the time of presentation. However, prognosis is believed to improve with earlier diagnosis [8]. Therefore, early diagnosis and treatment of sinonasal lymphoma are crucial, especially in patients of Asian descent, whose prognosis and outcomes have been reported to be more dismal compared to Western populations [9].

Conclusion

This case underscores the significance of sound clinical judgment and a heightened suspicion of malignancy when confronted with atypical symptoms and signs. In the context of a middle-aged patient of Asian origin, the consideration of extra nodal NK/T cell lymphoma nasal type becomes crucial when presented with atypical rhinosinusitis symptoms, with or without accompanying constitutional symptoms.

Acknowledgement

The authors would like to thank the patient for his permission and cooperation in writing this case.

Conflict of interest

None to declare.

Patients' consent for the use of images and content for publication

The patient provided verbal consent prior to the use of the mass and histopathological images, and the case for publication. Following the patient's passing, verbal consent was also obtained from his wife.

What is new in this case report compared to the previous literature?

- This case report highlights the importance of a high index of suspicion in at-risk person with atypical presentation of rhinosinusitis.
- Delayed recognition of atypical presentation of rhinosinusitis will lead to unnecessary management and should not be missed in clinical settings.

What is the implication to patients?

A delay in establishing the diagnosis of extra nodal NK/T cell lymphoma nasal type can lead to severe complications for patients due to inappropriate treatment. Despite the poor prognosis, it can be improved if the disease can be detected earlier.

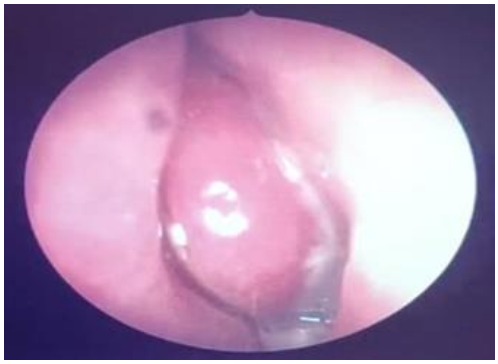
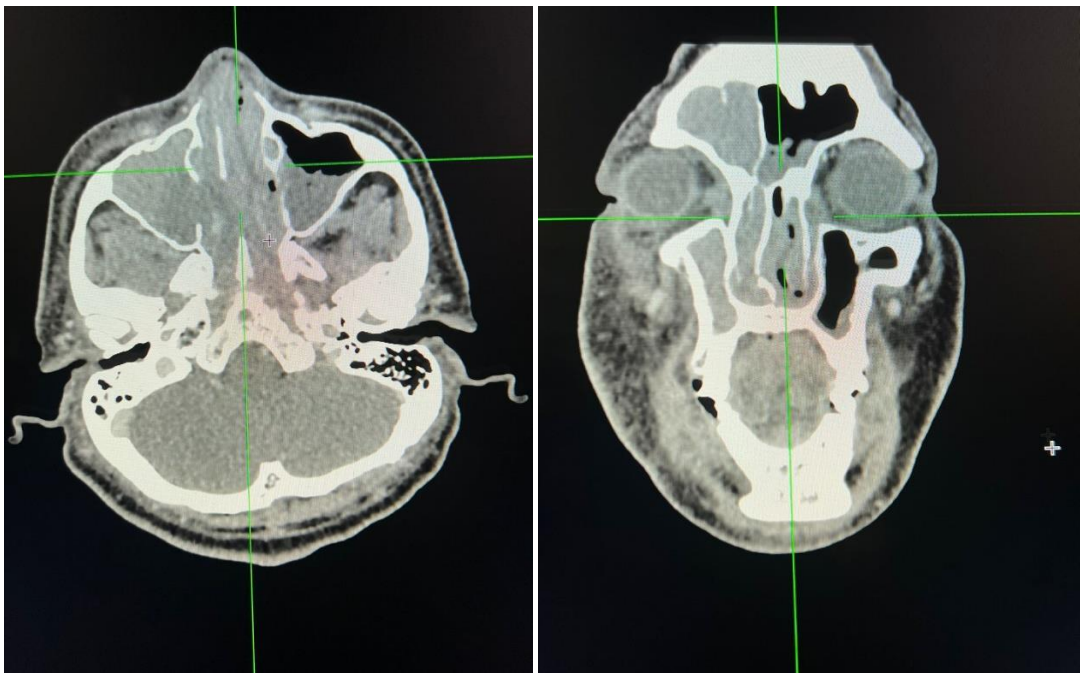
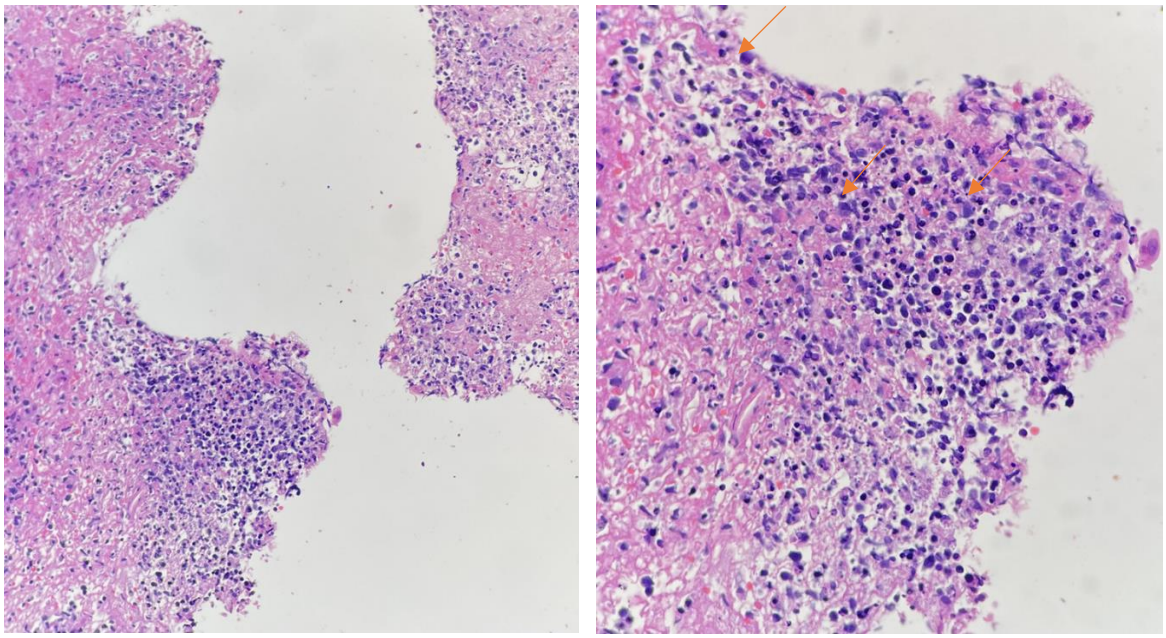


Figure 1. Photograph taken during right rigid nasal endoscopy reveal a mass of uncertain origin in the right nasal cavity.



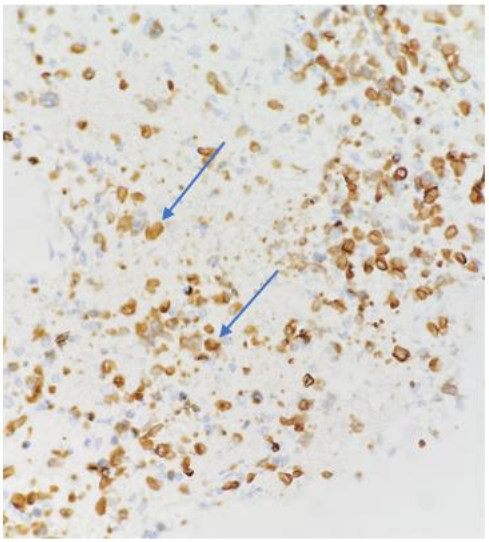
2(a) 2(b)

Figure 2(a) and 2(b). Axial and coronal view CECT of head showing bilateral nasal cavity filled with soft tissue density/fluid (right > left) resulting in severe narrowing.

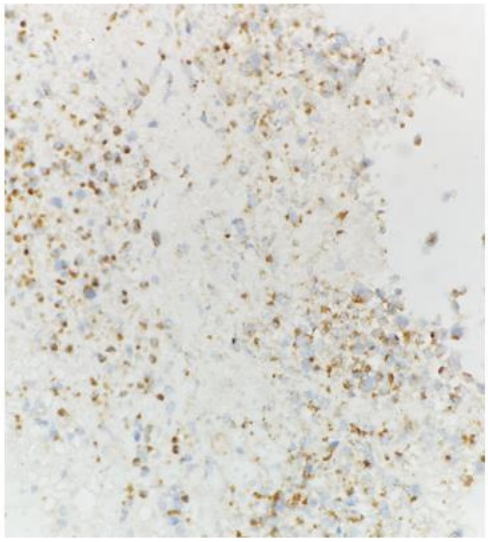


3(a) 3(b)

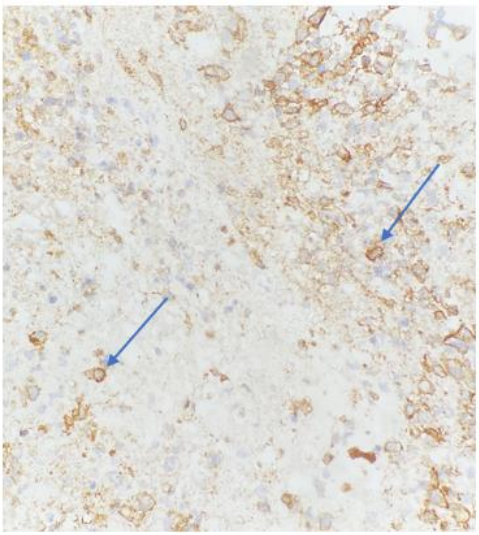
Figure 3(a) Hematoxylin and Eosin staining at 200X show predominantly necrotic tissue with few atypical lymphoid cells observed. 3(b) Hematoxylin and Eosin staining at 400X show medium to large sized cells exhibiting moderate cytoplasm, irregularly folded nuclei, coarse chromatin with inconspicuous nucleoli.



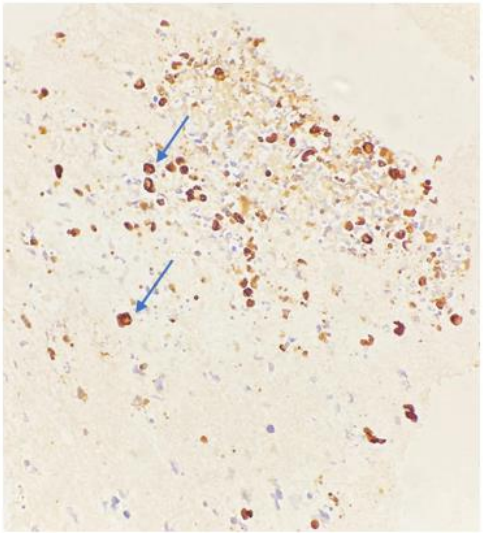
(a) CD3 at 400X Objective



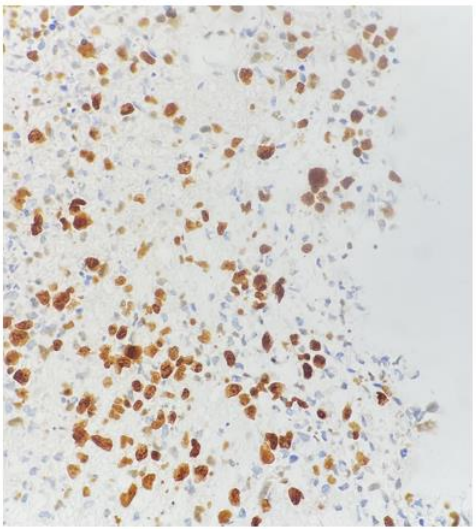
(b) TIA at 400X Objective



(c) CD56 at 400X Objective



(d) EBER-ISH at 400X Objective



e) Ki67 proliferative index of 60% at 400X objective

Fig. 4 (a – e) Immunohistochemical positive for CD3, TIA, CD56, EBER-ISH and Ki67

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