Peripheral natural killer (NK)/T-cell lymphomas constitute 10% to 15% of all non-Hodgkin’s lymphoma, with nasal lymphoma being the most common subtype. The nasal-type NK/T-cell lymphoma is an extranodal lymphoma of the nasal cavity and nasopharynx. It presents as a midfacial ulceration resulting in progressive facial destruction with functional and aesthetic deformity. It has a high prevalence among males in Asia and America. However, it is rare in the Western world, amounting to around 1% of the population. Frequently it presents in the fourth decade of life. An unusual presentation of nasal NK/T-cell lymphoma in an elderly Caucasian man originating as a nonmidline lesion in the nasal ala is presented in this case report. Approval was obtained by the Malta Health Ethics Committee.

Case Report

A 72-year-old Maltese man of Irish and Welsh origin with a history of interstitial nephritis was referred to the otolaryngology department with a 1-month history of crusting and erythema of the right nasal ala and a 1-year history of regular epistaxis. He was administered 3 courses of oral and topical antibiotics without improvement. On anterior rhinoscopy, an ulcerating crusted mass was noted arising from the right nasal ala, which was associated with swelling of the right infraorbital area. The nasal septum was completely normal. Neck examination and flexible nasoendoscopy findings were unremarkable. A biopsy of the ulcerating mass was taken under local anaesthesia. The histologic features and immunoprofile were consistent with high-grade peripheral T-cell lymphoma. It showed large atypical lymphocytes with large nuclei and prominent nucleoli. Mitotic figures and apoptotic bodies were present with areas of necrosis prominent. Immunohistochemistry showed diffuse expression of CD2, weaker expression of CD56, less diffuse expression of CD30, and very focal expression of CD3. A minority of the neoplastic lymphocytes expressed CD8. Flow cytometry confirmed the loss of expression of CD3, CD5, CD7, and CD43, with neoplastic cells expressing CD8 and HLA-DR and with weak expression of CD2, CD56, and CD16—consistent with extranodal NK/T-cell lymphoma. Polymerase chain reaction of serum Epstein-Barr antigen, which tends to be associated with extranodal NK/T-cell lymphoma, was positive with 2564 cp/mL.

The patient was referred for further treatment to the hematology and oncology departments. Magnetic resonance imaging of the head and neck was performed, which showed soft tissue thickening overlying the right nasal bone, in keeping with the lymphomatous process without extension to ipsilateral orbits. The bilateral turbinates and nasal septum were intact with no cervical lymphadenopathy. Staging bone marrow biopsy showed reactive features, and positron emission tomography–computed tomography scan outcome

Keywords

nasal NK/T-cell lymphoma, atypical, tumor

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Figure 1. Nasal natural killer/T-cell lymphoma involving nasal ala sparing the nasal septum (A) before and (B) after treatment.

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was unremarkable, confirming the localized nature of NK/T-cell lymphoma of the nasal ala. He was prescribed prednisolone and levofloxacin and scheduled for combined radiotherapy and chemotherapy. Given the history of interstitial nephritis and decreased renal function, intensive chemotherapy was abandoned.

**Discussion**

This is the first reported case in Europe of nasal NK/T-cell lymphoma presenting as a nonmidline nasal ala lesion in a Caucasian elderly man. This case is atypical in view of the patient’s age and presentation at the nasal ala with an intact nasal septum.

Nasal NK/T-cell lymphoma is a rare but aggressive, locally destructive necrotic lesion in the midline of the face. It may present as nasal stuffiness, chronic nasal obstruction, or purulent nasal discharge: all nonspecific clinical signs at presentation, making it difficult to diagnose early. Diagnosis rests on epidemiologic, clinical, and histopathologic characteristics. The crusting and necrotic surface of the lesion makes punch biopsy difficult to obtain. Hence, excisional or deep tissue biopsy needs to be taken. Histologic features of NK/T-cell lymphoma include angiocentric and angiodestructive growth with zonal necrosis and dense, dermal, atypical lymphoid cells. Immunohistochemistry aids to differentiate NK/T-cell lymphoma from other lymphoproliferative malignancies, as opposed to computed tomography scan or magnetic resonance imaging, which shows nonspecific features.1,2

Treatment includes radiotherapy for all patients with local disease and/or CHOP chemotherapy (cyclophosphamide, doxorubicin, vincristine, and prednisolone). Despite this being effective, NK/T-cell lymphoma has a high recurrence rate, reaching 50%. It is associated with a high mortality if not managed timely, resulting in 5-year survival rates of 70% and 30% in the early and late stages of disease, respectively.1,4

**Conclusion**

The nonspecific presentation of NK/T-cell lymphoma results in delay in its diagnosis, adversely affecting disease prognosis. High clinical suspicion is needed for facial ulcerations that fail to respond to conventional medical therapy even if these present as nonmidline lesions, as demonstrated in this case.

**Author Contributions**

Svetlana Doris Brincat, manuscript design, draft, and revision; Alison Abdilla, manuscript design and revision; Hermann Karl Borg Xuereb, manuscript design and revision. All authors have contributed significantly to this publication, are aware of the submission, and agree with it.

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**Patient Confidentiality**

Although the photograph does not reveal the person’s identity, the patient has granted the authors the right to publish the photograph online and in print in a scientific medical journal.

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