CORRESPONDENCE

Two Cases of NK/T Cell Lymphoma Presenting as Chronic Rhinosinusitis

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Abstract T/NK cell lymphoma is a rare subtype of lymphoma that is known to be of aggressive nature. We present two cases of this disease that originally presented with symptoms consistent with chronic rhinosinusitis. We outline how we managed the diseases and review literature on it.

Keywords Non hodgkins lymphoma · Chronic rhinosinusitis · Plastic surgery · NK cells

Introduction

T/NK cell lymphoma is a rare subtype of lymphoma. It causes necrosis and can be found anywhere in the body but has a predilection for the gastrointestinal tract, oral/nasal cavity, skin and the testis. It is commonly found in the nasal and paranasal region [1]. We present two patients of diagnosed T/NK cell lymphoma who presented with a nasal polyp and developed bony destruction of the nose and adjacent facial bones and soft tissue.

Case

Case 1

In 2012, a 30-year-old African lady was evaluated for chronic rhinosinusitis for 6 months. She underwent polypectomy, biopsy of which showed T/NK cell lymphoma on histopathological examination. She was given chemotherapy using CHOP regimen. After receiving 6 cycles of chemotherapy, she developed blackening of the nose, which led to an ulcero-proliferative mass causing destruction of the upper lip, entire hard palate, including the entire maxillary alveolus except left maxillary molars, nose, soft tissue around the nasal bones, turbinates and nasal septum (Fig. 1a). This was suggestive of a local relapse. The patient received supportive treatment. PET showed disease localized to the primary lesion. She underwent debridement, and after negative biopsies from the healed edges of the defect, reconstruction by Osseointegrated silicon of the soft tissue defect of the face (Fig. 1b) facilitating her feeding, as well as restoring her appearance. Following this, she went to her parent country.

Case 2

In 2014, a 29-year-old Indian male had a similar history for 2 years, and underwent treatment outside the hospital. He underwent functional endoscopic sinus surgery with turbinectomy and septoplasty. Histopathology showed an inflammatory polyp. The patient developed progressive blackening of his nose. CECT showed an ulceroproliferative lesion destroying the nose, turbinates, and maxilla. Histopathology was reviewed and it confirmed nasal T/NK cell lymphoma, which was CD3, CD5, CD2, and granzyme B positive. He received 2 cycles of chemotherapy with

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Fig. 1 **a** Before plastic reconstruction, after debridement. **b** After plastic reconstruction

SMILE regime, after which he received IGRT. He was in continuous care of a plastic surgeon. He then received four more cycles of chemotherapy. Patient tolerated the treatment well and was discharged. He has been advised plastic surgery for his facial defect and is on regular follow up.

Discussion

These two cases show extra-nodal T/NK cell lymphoma presenting with chronic rhinosinusitis. The diagnosis was not made until the biopsy was reviewed. Thus, a high index of suspicion is needed for the diagnosis. Even though this disease is uncommon in India, but with international tourism, we should consider it as a differential diagnosis. The disease is clustered in East Asia, Africa and the Latin Americas.

Our experience with an l-asparaginase based regimen combined with radiotherapy was better than conventional chemotherapy with CHOP. The need for local reconstructive surgery should be recognized. In late presentations, the involvement of a plastic surgeon early may be necessary.

NK cells are cytotoxic cells that are target and lyse virus-infected and tumorigenic cells [2]. They express CD2, CD7 and CD8 because of a common ontogeny with T cells, and characteristically express CD56 and CD57, which are “NK lineage associated markers” [3]. Sometimes, CD3 positivity may be found along with NK cell markers, which is why WHO classified them as a common entity of NK/T cell lymphoma [4].

NK/T cell lymphoma is subdivided into nasal, non-nasal lymphoma, and aggressive leukemia [4]. The nasal lesion is a destructive mass lesion involving the nasal cavity, nasopharynx, paranasal sinuses, tonsils, hypopharynx, and larynx. Destruction of hard palate leads to a midline perforation. Non-nasal lymphoma involves any site. Interestingly,

non-nasal NK/T cell lymphoma is commonly found in places nasal NK/T cell lymphoma metastasizes to, i.e. skin, gastrointestinal tract, salivary gland, spleen and testis. Therefore, a non-nasal disease warrants a panendoscopy and imaging to exclude an occult disease. NK cell leukemia is an aggressive form of the disease, which presents with high fever, weight loss, jaundice, skin infiltration, lymphadenopathy and hepatosplenomegaly.

A patient of T/NK cell lymphoma should be thoroughly evaluated with a history and physical examination. An extensive biopsy of the lesion should be done, as a small one may only show necrosis. An unfixed biopsy is preferable as a flow-cytometry can be done which may show NK cell specific markers. If a fresh biopsy is unavailable, then NK cells and T cells need to be differentiated with TCR rearrangement studies. CT and MRI may be done to see the local involvement. PET scan is done to see distant metastasis and helps with radiation planning [4].

Ebstein Barr Virus (EBV) DNA is found because of the viral DNA release from apoptosis of tumor. EBV DNA quantification is correlated with disease control [3]. And it is a prerequisite for the diagnosis according to the WHO classification [3, 5]. The disease staging by the Ann Arbor method is not ideal as the disease is primarily extranodal in presentation. A staging criterion, International Prognostic Index (IPI) may be better as it can also be used for prognostication [6].

The treatment of choice for stage I/II nasal T/NK cell lymphoma is combination of chemotherapy and radiotherapy [7]. It is curative in 70–80 % of all stage I/II nasal T/NK cell lymphoma. Chemotherapy is usually non-anthracycline based and early radiotherapy is preferred (concurrent or sequential). In advanced disease, chemotherapy is the mainstay and CHOP regimen is unsatisfactory, and SMILE regimen, with dexamethasone, methotrexate, ifosfamide, l-asparaginase and etoposide, seems promising [6].

Only chemotherapy is the mainstay of treatment of non-nasal T/NK cell lymphoma. NK cell leukemia is very rare and treatment is with L-asparaginase containing regimens. HSCT may lead to improved outcomes but prospective studies are unavailable [4].

Conclusion

A high index of suspicion is needed for the diagnosis of T/NK lymphoma in patients of chronic rhinosinusitis. Even though the disease rare in India, it should be considered as a differential. With appropriate chemotherapy, the disease is imminently curable. The need for local reconstructive surgery with prosthesis to repair lost tissue and rehabilitate the patient should be recognized. In aggressive cases or late presentations, the involvement of a plastic surgeon early may be necessary.

Compliance with Ethical Standards

Conflict of interest None.

Ethical Approval This article does not contain any studies with human participants performed by any of the authors.

Informed Consent Informed consent was obtained from all individual participants included in the study.

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