Nasal Natural Killer/T Cell Lymphoma

Milind Anil Bhatkule · Manjusha Shripad Dhawle ·
N. R. Kumbhakarna · Rajan S. Bindu

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Abstract Nasal natural killer/T cell lymphomas are locally destructive mid-facial necrotizing lesions. It is an uncommon disease but, usually shows a highly aggressive clinical course. Amongst the sino-nasal lymphomas, it is the most common type. The non-specific clinical features constitute a major stumbling block in the early diagnosis and management of these lymphomas. We report here a case of nasal NK/T cell lymphoma in a 23-year old male with review of literature.

Keywords Lymphoma · Nasal · Natural killer

Introduction

Natural killer NK/T cell lymphoma is the most common malignant lymphoma among the sino-nasal lymphoma. NK/T cell lymphomas are categorized as either nasal or extra-nasal in origin. They are aggressive, locally destructive, midline facial necrotizing lesions. Initially these lesions were labeled as lethal midline granulomas [1–3], a term which is slowly being phased out. The non specific clinical symptoms constitute a major stumbling block in the early diagnosis and management of these lymphomas. Thus, we need to be aware of the uncommon presentation of this lymphoma. We report here a case of nasal NK/T cell lymphoma in an apparently healthy male that progressed rapidly in a short span of time

Case Report

A 23-year old male presented with swelling and erythema over nose (Fig. 1). There was history of nasal stuffiness. There was no history of any kind of nasal discharge. Systemic examination revealed cervical lymphadenopathy. Routine hematological and biochemical investigations were normal. CT scan showed enhancing soft tissue density lesion in the nasal cavity with destruction of anterior part of nasal septum. Right maxillary region showed soft tissue thickening. A biopsy was performed. Histopathological examination showed diffusely scattered large cells with large hyperchromatic pleomorphic nuclei with angio-centric distribution of cells. Large areas of necrosis were seen (Fig. 2). Immunohistochemistry was positive for LCA (leucocyte common antigen), CD3, and CD56.

Thus, a diagnosis of nasal NK/T cell lymphoma was reached.

Discussion

Extra nodal NK/T cell lymphoma, nasal type, accounts for 7–10 % of all non-Hodgkin lymphomas in Asia and Latin America, but for only 1 % in Europe and North America.

The presence of this disease in young individuals in South India has been reported [5]. Nasal NK/T cell lymphoma is an uncommon disease, usually showing a highly aggressive clinical course. According to WHO classification, extra-nodal nasal natural killer cell/T cell (NK/T cell) lymphoma is derived from natural killer cells and/or cytotoxic T lymphocytes. It typically involves the nasal cavity and the para-nasal sinuses [4]. Mature NK/T cell lymphomas account for only 10–15 % of non-Hodgkin lymphomas. Amongst them, the most common are the

M. A. Bhatkule · M. S. Dhawle (✉) · N. R. Kumbhakarna ·
R. S. Bindu
Department of Pathology, Government Medical College,
Aurangabad 431001, Maharashtra, India
e-mail: drmilindbhatkule@yahoo.co.in

Fig. 1 Male presented with swelling and erythema over nose

Fig. 2 Histopathological examination showed diffusely scattered large cells with large hyperchromatic pleomorphic nuclei with angio-centric distribution of cells

nasal NK/T cell lymphomas. Terms like non-healing granuloma, malignant granuloma and lethal midline granuloma have been used to describe NK/T cell lymphoma lesions [6].

Apart from nasal cavity, other sites of involvement by NK/T cell lymphoma are skin, gut, testis, kidneys, upper respiratory tract and rarely, the eyes [7]. Skin is the most common site for NK/T cell lymphoma after nasal cavity.

Evidence of EBV has also been found in NK/T cell lymphoma [7]. They are rare and are mainly seen in Asia and Latin America. Elderly males in the age group of 50 years and above are affected.

The most common presentation is chronic nasal obstruction or purulent rhinorrhoea [1]. It is important to note the non-descript nature of these presenting symptoms, which may result in delayed diagnosis. Systemic symptoms like fever and weight loss are noticed in advanced cases [1]. Our patient presented with nasal obstruction.

Histologically, NK/T cell lymphoma shows a mixed cellular infiltration consisting of atypical lymphocytes, plasma cells, eosinophils and histiocytes. Angiocentricity and angio-invasion is generally seen. Our case showed large number of atypical lymphocytes.

NK/T cell lymphomas show a positive immune reaction for CD45Ro, CD3 and CD56, thus, demonstrating presence of T cell and natural killer cell markers. Negativity for b cell markers like CD30 And CD 20. In our case, these markers were demonstrated immunohistochemically, confirming the diagnosis of NK/T cell lymphoma.

NK/T cell lymphoma follows a rapid and aggressive course. Prognosis is especially poor, when there is systemic involvement. The median overall survival rate is 12.5 months. Better prognosis can sometimes be achieved with early and accurate diagnosis and aggressive chemotherapy and radiotherapy [6].

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