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Testicular Metastasis ofPrimary NK / T Nasal Type Lymphoma:A Case Report

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Abstract

Testicular lymphoma is an aggressive disease with a very poor prognosis. Nasal-type natural killer/T-cell lymphoma (NKTCL-N) in particular is very uncommon and has a rapidly progressive fatal course. We report a case of a young patient who presented a primary NK/T cell nasal type lymphoma with a secondary testicular location.

Keywords: Testicular metastasis; Nasal-type natural killer/T-cell lymphoma

Abbreviations: NK: Natural Killer; NKTCL-N: Natural Killer T Cell Nasal Lymphoma

Introduction

There are three categories of natural killer cell tumors: extranodal NK/T-cell lymphoma, the nasal type (NKTCL-N), an aggressive NK cell leukemia, and blastic NK cell lymphoma. The NKTCL-N lymphoma has two distinct variants, depending on the site of origin (nasal versus non nasal) [1]. The Nasal variant affects mainly the midline structures including the nasal cavity, nasopharynx, and paranasal sinuses, whereas the extranasal one involves the skin, the gastrointestinal tract, the salivary glands, the spleen, the lungs, and the testis [1]. The World Health Organization (WHO) classification groups both nasal and extra nasal NKTCL-N lymphoma in the same category as "nasal type" although they have different clinical manifestations, treatment approaches, and prognosis [2,3]. Testicular NKTCL-N lymphoma is a very rare entity with an aggressive natural history with tumors cells expressing CD56. In literature, there are about 16 cases of primary NKTCL-N lymphoma, Yet, this article will focus on a double presentation of this rare lymphoma.

Case Report

This is a 28 year old patient without specific medical history admitted to internal medicine department for the treatment of the right nasal cavity tumor (Figure1). He received a nasal endoscopy with biopsy. The immunohistochemical study showed a lymphoma T / NK nasal type. An 18f-FDG PET-SCAN used for staging showed a very intense active process of right nasal cavity extending to the posterior ethmoid sinuses associated with an ipsilateral lymph node active involvement at the right mandibular (Figure 2). This test also showed a left testicular hypertrophy with intense pathological active foci (Figure 3). Testicular tumor markers were negative (bHCG, AFP and LDH). The results of a bone marrow biopsy for his pancytopenia found no anomaly. The patient underwent a left orchiectomy with pathological results in favor of a seminoma. A second review of slides and an additional immunohistochemistry showed a secondary location of a T / NK nasal type testicular lymphoma. The profile is positive anti Ki67 antibody (90%), positive anti-CD3 antibodies, positive anti CD5 antibody and positive anti CD56 antibodies (Figure 4,5). During his hospitalization, the patient presented fever, chills, swelling of the face and nasal obstruction on the right side. The patient received vancomycin for 10 days without clinical or biological improvement. Chemotherapy combining cyclophosphamide, doxorubicin, vincristine, and a steroid bolus were administered, but the patient developed a deterioration of respiratory state due to hypoxic lung disease, whose evolution was fatal.

Discussion

Lymphomas of the nasal cavity are very rare and represent less than 1% of all malignant tumours of head and neck. The nasal area

is the usual site, but the whole aero-digestive tract can be affected [4,5]. The clinical symptoms are nonspecific making the diagnosis difficult. Patients may present epistaxis, nasal obstruction, ptosis, and facial or dental neuralgia. In our case, the first sign was a tumor in the nasal region. The extension to neighbouring tissues and the existence of bone erosion are very suggestive but not specific to diagnosis [6,7]. Skin and subcutaneous tissues are the be most common metastatic sites but the gastrointestinal tract, nervous system and testicles can be affected as in our case [8]. To our knowledge no secondary testicular location, as observed in our case, has been reported in literature. A bifocal presentation of Primary Testicular Extranasal NK/T-Cell Lymphoma was described at Clemenceau Medical Center in Beirut [9]. (The inverse of our case). The association of EBV infection contributes to the increased risk, which adversely affects the patient survival [10-12], suggesting that the presence of EBV may correlate closely with prognosis in primary NKTCL-N lymphoma of the testis. Despite intensive chemotherapy treatments, this disease is highly aggressive with an early skin, aero-digestive tract, or soft tissue involvement.

Figure 1: Anterior view of right nasal cavity tumor .

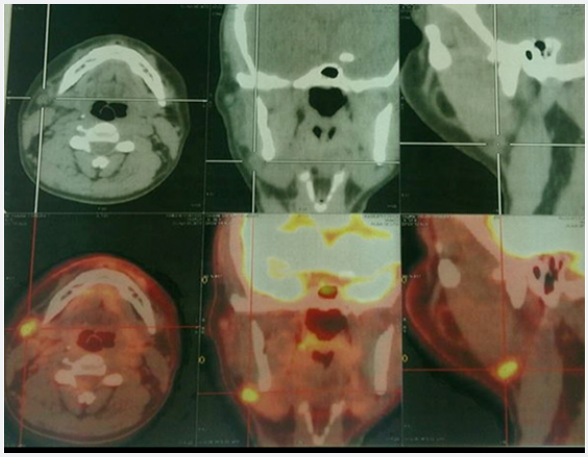

Figure 2: A CT of sinuses showing the right nasal mass.

Figure 3: A left testicular hypertrophy with intense pathological active foci.

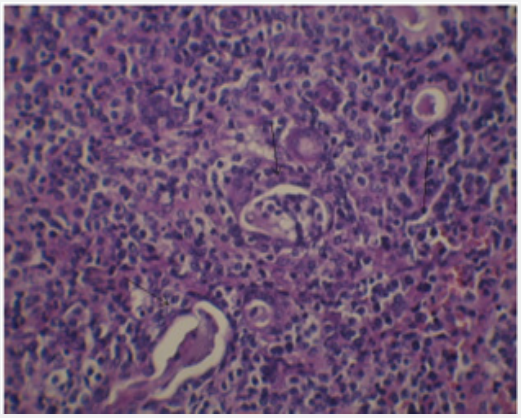

Figure 4: Dense and diffuse polymorphous population of tumor cells consisting of small to medium sized and large cells with irregular nuclei.

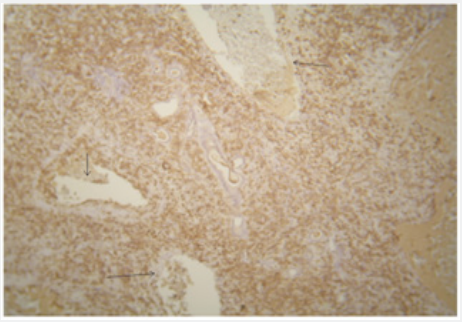

Figure 5: An immunohistochemistry with tumor cells expressing CD56.

Conclusion

The N.K. / T-cell nasal type lymphoma is a rapidly spreading and invasive tumour, rarely associated with testicular metastasis.

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Though diagnosis is difficult, the immunohistochemistry remains a very useful tool. The treatment is based mainly on chemotherapy.

Conflit of interest

All authors report no conflict of interests

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