A nasal swelling revealing a metastatic testicular extranodal NK/T-cell lymphoma: A case report and a review of literature

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A B S T R A C T

Testicular ENKTCL is a rare disease. Asia is the most affected. Primary testicular NK/T-cell lymphoma is rare. Metastases are early and the prognosis is poor. Metastases mainly involve the lymph nodes, skin, contralateral testis, bone marrow, spleen and central nervous system. Nasal metastasis giving rise to bifocal presentation is extremely rare. We report the management of a patient initially seen for a nasal swelling with a hidden history of scrotal swelling, in whom nasal biopsies as well as the analysis of the orchidectomy part made it possible to retain the diagnosis of ENKTCL of the testis with a nasal metastasis.

Introduction

Extranodal nasal type natural killer/T-cell lymphoma of testis is a very rare disease. Metastases and non-testicular locations are usually the lymph nodes, skin, bone marrow, spleen, adrenals, and central nervous system; nasal metastasis giving rise to bifocal presentation is extremely rare. The disease is characterized by a strong metastatic potential and recurrence even when it is discovered early; as well as a very poor prognosis, making it an extremely serious condition. Even the most aggressive treatment (Orchidectomy + Chemotherapy ± Radiotherapy) does not seem to produce satisfactory results.

Most published cases are concentrated in Asia. To our knowledge, no Moroccan case has been published to date. We report the management of a patient who was initially seen for a nasal swelling with a hidden history of scrotal swelling, in whom nasal biopsies as well as the analysis of the orchidectomy part made it possible to retain the diagnosis of extranodal nasal type natural killer/T-cell lymphoma of testis with a nasal metastasis.

Case report

A 28 Arab male Moroccan, with no comorbidities known in general and lymphoma in particular. His family history was unremarkable for cancers in any first- or second-degree relatives.

The patient had initially consulted for a right nasal swelling reaching the eyelid and taking the upper lip (Fig. 1) all evolving in a context of fever, night sweat and weight loss estimated at 10 Kg in 2 months. A thorough questioning and clinical examination revealed a painless left scrotal swelling of 7.4 × 7 * 6 cm which had preceded by 2 months the nasal swelling, for which he had taken antibiotics for self-medication.

A cerebral and sinus MRI was performed showing a complete filling of the right maxillary sinus prolapsed through an accessory ostium to the homolateral nasal cavity completely filling the choane evoking a Killian polyp.

A testicular ultrasound showed an enlarged left testicle, site of 3 hypoechoic nodular lesions. The usual markers for testicular cancer were all negative (APF, HCG, LDH, CA 19-9 and CEA).

Endoscopic biopsies of the nasal lesion were performed. The anatomopathological study as well as the immunostaining made it possible to diagnose a nasal extranodal NK/T-cell lymphoma (ENKTCL). A PET-CT was performed and showed a very intense active process of the right nasal fossa extending to the level of the posterior homolateral ethmoid sinus as well as a left testicular enlargement with intense pathological active foci (Fig. 2). The patient underwent a left orchidectomy. The anatomopathological study was in favor of an extranodal NK/T-cell lymphoma of the testis. The tumor stained positive for CD 56, CD 3 ε, Granzym B, TIA-1 and EBER; and negative for CD 7, CD 20 and CD 8 (Fig. 3). The patient also had positive EBV tests.

The protocol (Dexamethasone: 40 mg/m2 the 2nd et 4th day, Methotrexate: 2mg/m2 the first day, Ifosfamide: 1,5g/m2 the 2nd et 4th day, L-asparaginase: 6000 U/m2 the 8th, 10th, 12th, 14th, 16th, 18th day, Etoposide: 100mg/m2 the 2nd et 4th day) combined with radiotherapy (50 Gy in total).

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While under chemotherapy, 03 months after the onset of scrotal swelling and 01 month after diagnosis, the patient was admitted to intensive care where he unfortunately died of generalized sepsis.

Discussion

Extranodal NK/T-cell lymphoma (ENKTCL) is divided into two subgroups depending on the primary site: nasal and extranasal. The Nasal type affects the nose, nasopharynx and upper aerodigestive system. The extranasal type affects the skin, the gastrointestinal tract, the testicle and other sites. The association with EBV infection is almost constant.1

Classically, at immunohistochemistry, NK cells express CD 2, cytoplasmic CD3 epsilon (but not surface CD 3), CD 16, CD 56, CD 94 and cytotoxic molecules (Perforin granzyme, TIA-1).2 Huang et al.3 reported an abnormal expression of CD 20 in 10.3% of cases which could be explained by either the transformation of a rare type of cell expressing both CD 20 and the usual markers of NK cells, either by an aberrant antigenic expression due to the neoplastic context. The expression of CD 20 could be synonymous with greater proliferation potential and greater aggressiveness. Testicular NK/T-cell lymphoma is rare. It is called primary testicular NK/T-cell lymphoma when the testicle is the only organ affected.4

Asia is the most affected region in the world, followed by Indigenous Americans from Mexico. The clinical presentation of primary testicular NK/T-cell lymphoma is that of a large unilateral bursa evolving in a context of fever, night sweats and weight loss.

Despite the aggressive approach (orchidectomy ± chemotherapy± radiotherapy) practiced by the different teams, the results are not satisfactory. In a meta-analysis of 39 primary testicular NK/T-cell lymphomas, Huang et al.5 reported that 80% of patients metastasized after 06 months of follow-up and that survival at 2 years was 34% with a median survival at 15.3 months. The median age in Huang’s series was 45. The most common metastatic sites are the lymph nodes, skin, contralateral testis, bone marrow, spleen and central nervous system. In a series of 31 cases, Liang et al.5 reported a median survival of 5.8 months for primary testicular NK/T-cell lymphomas and 4.5 months for non-primary cases.

The nasal sinuses are an extremely rare secondary location. Huang et al.3 do not report nasal metastases. Liang et al. in a series of 31 cases of testicular NK/T -cell lymphomas reported a case of nasal metastasis among the 15 primary testicular NK/T-cell lymphoma cases and 2 cases of nasal involvement in the group of 16 non-primary testicular NK/T-cell lymphomas.

Conclusion

Testicular natural killer/T-cell lymphoma is a rare disease. Primary testicular NK/T-cell lymphoma is very rare. Nasal metastasis is
extremely rare. The high metastatic potential as well as the low survival rate despite aggressive management make it an extremely serious disease. There is no standard protocol for the management of the disease. Progresses in targeted therapy are to be hoped for.

Author contributions

Dr. CA Tombet and A. El Houmaidi conducted the study and wrote the manuscript. Drs. M Aynaou, T Mhanna and M Chennoufi participated in the care of the patient. Teacher A Barki supervised the writing.

Consent

Written informed consent was obtained from the patient’s next of kin for publication of this case report.

Declaration of competing interest

The authors do not declare any conflict of interest.

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Fig. 3. A-Dense and diffuse polymorphous population of tumor cells consisting of small to medium sized and large cells with irregular nuclei. B- An immunohistochemistry with tumor cells expressing CD56.