CASE REPORT

Sinonasal NK/T-cell lymphoma

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Summary
Introduction: Nasal NK/T-cell lymphoma is a rare but well-known clinical entity. Thanks to improvements in anatomopathology and the development of routine immunohistochemistry, the diagnosis of nasal NK/T-cell lymphoma (NK for “natural killer”) was recognized by the WHO in 2001. The main differential diagnosis is with Wegener’s granulomatosis. Treatment is based on radiotherapy and chemotherapy. Prognosis is poor, with variable evolution and sometimes rapid progression.

Case report: The authors report the case of a 54-year-old man without known pathological history, who presented with bilateral nasal obstruction with purulent rhinorrhea. Diagnosis was made on the basis of immunohistochemical study of biopsy samples. The tumor was graded IE on the Ann Arbor classification. Treatment, comprising CHOP chemotherapy followed by radiotherapy, achieved total remission at 8 months’ follow-up.

Discussion/Conclusion: Sinonasal NK/T-cell lymphoma is rare. Diagnosis is based on immunophenotypic and molecular characteristics. It is an aggressive lymphoma, requiring multidisciplinary management. Prognosis is poor.

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Introduction

Nasal NK/T-cell lymphoma, formerly known as malignant centrofacial granuloma, is a rare clinicopathologic entity characterized by a necrotic process originating in the nasal cavity and extending to the medio-facial bone structures with centrifugal destruction of the facial bone. The advent of immunohistochemistry enabled its recognition by the WHO in 2001. Evolution is spontaneously fatal. Treatment is based on radiotherapy and chemotherapy.

Sinonasal locations are rare. We report one such case and discuss the specificities of this location.

Observation

A 54-year-old man without known pathological history presented with bilateral nasal obstruction of progressive onset, associated with fetid bilateral purulent rhinorrhea and upper dysphagia of 5 months’ evolution in a context of apyrexia and 4 kg weight-loss over 3 months.

ENT examination found a fleshy burgeoning polypoid formation, bleeding on contact and filling the entire right nasal cavity, pushing back the ipsilateral middle turbinate and nasal septum. Endoscopy was impossible due to the size of the mass. Oral cavity examination found hard palate ulceration (Fig. 1). Otherwise, clinical examination, notably of the lymph-node regions, was normal.

Bacteriological examination of the secretions, sampled by direct swabbing of the nasal cavities, was negative. Sinonasal CT found filling of the maxillary sinuses and right
nasal cavity with lysis of the medial wall of the right maxillary sinus and floor of the right nasal cavity (Fig. 2a and b).

Histopathologic analysis of biopsy samples from the right nasal cavity and hard palate, taken under general anesthesia, found friable tissue with necrotic areas, suggestive of infectious or tumoral pathology, without being able to decide between the two. Immunohistochemistry diagnosed sinonasal NK/T-cell lymphoma. The lymphoid elements were positive for cytoplasmic CD3, negative for pan-cytokeratin and positive for CD56 (Fig. 3).

Extension assessment, comprising chest X-ray, thoracic-abdominal-pelvic CT, digestive fibroscopy and osteomedullary biopsy, was negative. Biological analysis was normal, and TPHA-VDRL, HIV and EBV serology was negative. The lymphoma was graded IE on the Ann Arbor classification (Table 1).

Treatment consisted in adjuvant polychemotherapy, with five CHOP cycles (doxorubicin, vincristine, cyclophosphamide and prednisone), followed by locoregional external radiotherapy (paranasal sinus and nasal cavities: 48.6 GY in 27 sessions). At 8 months’ follow-up, the patient was in complete clinical and radiological remission.

Discussion

Sinonasal NK/T-cell lymphoma is rare, but more common in Asia, Mexico and South America than in Europe and North America [1]. There have been a few reports concerning this location [2–4]. Onset is usually during the 5th decade of life in Western countries [3,4]; in Asia, patients are younger, with a mean age of 40 years, and show male predominance (sex ratio, 2/3) [4,5].

Clinical presentation largely consists of local signs such as unilateral nasal obstruction, purulent and/or blood-streaked rhinorrhea, recurrent epistaxis or chronic sinusitis [4,6]. General signs, such as fever, weight-loss or night sweats, are rare and concern advanced stages. Signs of regional extension (jaw swelling or ophthalmic, auditory,
Sinonasal NK/T-cell lymphoma 147

pharyngeal or neurological disorder) may be the presenting symptoms [4].

Clinical examination generally finds an anterior ulceronecrotic lesion, bleeding on contact and filling the nasal cavity, usually located on the lateral wall [4,7]. Lymph-node involvement varies from series to series and never exceeds 25% of cases [4,8].

CT guides diagnosis, often finding an aspect suggestive of solid tumor with little if any contrast uptake. Bone structure destruction is found in less than half of cases and especially in case of large tumor.

MRI contributes to assessment of extension to adjacent structures, differentiating tumoral from inflammatory processes. Biological analysis systematically finds an inflammatory syndrome.

Diagnosis is confirmed by histologic and immunohistochemical biopsy analysis. Immunohistochemistry determines lymphoid element phenotype and is essential to diagnosis. NK etiology is confirmed from expression of CD2, CD56 and cytotoxicity markers (T1a, Granzyme B, perforin); CD5, CD4 and CD8 are negative; CD3 expression is variable (absence of superficial CD3 marker) [7]. Association with EBV is almost systematic. In the present case, only the immunohistochemical study provided certainty. There is a problem of differential diagnosis with respect, for example, to Wegener's granulomatosis [4].

Treatment for sinonasal NK/T-cell lymphoma is not as yet well codified, and depends mainly on the stage of the disease. Some teams generally use anthracycline-based polychemotherapy followed by external radiation consolidation therapy for patients younger than 60 years of age and the same association but without anthracycline in older patients [1]. Others still recommend radiation therapy alone in less advanced stages, as the failure rate in primary chemotherapy reaches 40%. Radiation therapy following failure of chemotherapy is associated with improved prognosis [9]. Overall survivorship for all treatment modalities taken together is around 37% [7].

Conclusion

Sinonasal locations for NK/T-cell lymphoma are rare. Diagnosis is founded on immunohistochemistry. Treatment associates chemo- and radiotherapy. Overall prognosis is poor, even with treatment. The major issue at present is to standardize treatment protocols in multidisciplinary teamwork between oncohematologists and ENT physicians.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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