Aggressive Nasal Extranodal Nk/t Cell Lymphoma: Case Report

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Abstract

Objective: Nasal extranodal NK/T-cell lymphoma is a rare aggressive lesion, characterized by a destructive process of the upper respiratory tract that has an unusual and rapid evolution. Diagnosis is often difficult and requires expert clinical examination and analysis of biopsies using immunohistochemistry.

Case report: A 34 years-old man with no relevant medical history, presented with an asymptomatic necrotizing lesion on palate and nasal obstruction. Two biopsies were performed with no conclusive results. By the third time, atypical lymphoid tissue was found surrounded by intense necrosis. The diagnosis was confirmed by immunohistochemistry. Chemotherapy was the treatment of choice follow by radiotherapy. The patient presented satisfactory evolution but one year after treatment a recidive was detected and patient died 5 months later from lung metastasis.

Conclusion: Suspicious ulcerative lesions in the midline head and neck region must have extranodal NK/T-cell lymphoma as differential diagnosis and repeated biopsies should be taken to confirm diagnosis.

Introduction

The extranodal NK/T-cell lymphoma is a rare type of non-Hodgkin lymphoma once known as lethal midline granuloma that sometimes can be mistaken for polymorphic reticulosis or lymphomatoid granulomatous, all of which have similar characteristics (Califano et al., 1998). The most common site is the upper respiratory tract, in addition to skin, gastrointestinal tract, spleen, breasts, testis and kidney (Cheung et al., 2002; Emile et al., 1996; Gourin et al., 2001; Hon et al., 2002; Mosqueda-Taylor et al., 1997). It is an idopathic, ulcer/necrotizing lesion with a strong inflammatory component, angiocentric and angiodestructive. The necrosis leads to destruction of structures and frequently to bone sequestration. The initial symptoms are rhinorrhea, nasal obstruction, epistaxis and in some cases, ophthalmic complications may precede the disease or appear in its course (Hon et al., 2002; Rodrigo et al., 2005). Systemic symptoms, such as fever and weight loss, are only present in advanced stages of the disease (Gourin et al., 2001). Histologically, is characterized by a polymorphic inflammatory cell infiltrate containing eosinophils, neutrophils, histiocytes and atypical lymphocytes (Califano et al., 1998). Due to the extensive necrosis and the inflammatory component, more than one biopsy may be necessary for the tumoral tissue to be identified. Immunohistochemistry can confirm the diagnosis by the T cell markers CD2, CD3 and CD7, as well as the NK cell marker CD56 (Rodrigo et al., 2005). In some cases, there are evidences of the Epstein-Barr virus, suggesting its involvement in the pathogenesis of the disease (Cheung et al., 2002; Emile et al., 1996; Hon et al., 2002; Mosqueda-Taylor et al., 1997). The extranodal NK/T-cell lymphoma has a poor prognosis, leading to death due to cachexia, hemorrhage or intercurrent infection (Lee et al., 2006).
lymphadenomegally was taking place of the jugular-carotid chain and accessory spines. Chemotherapy was chosen for initial treatment, followed by radiotherapy (Fig.3). The patient responded well to the treatment. After 2 years of follow-up, a maxillary reconstructive surgery was planned. During pre-operative exams, a recidive was found and patient died 5 months later from tumor complications.

Discussion

The extranodal NK/T-cell lymphoma is a rare lesion with a higher incidence in Asiatic countries (Rodrigo et al., 2005). It usually presents a poor quantity of atypical lymphocytes, and the extensive areas of necrosis make it difficult to choose the best site for biopsy. The common result of the histopathological diagnosis is chronic and acute inflammation. Necrosis favors the entrance of infectious processes that can lead to sepsis. Differential diagnosis can also be confusing since symptoms as secretion, nasal obstruction and ulceration may characterize other pathologies as well, such as Wegener’s granulomatosis (WG), blastomycosis, tuberculosis, adenocarcinoma, squamous cell carcinoma, nasal destruction for cocaine abuse (Rodrigo et al., 2005), among others. Immunohistochemistry is an important tool to confirm diagnosis, with positive expression for CD2, CD3 and CD56. Altemani et al. (2002), in a study about the characteristics of nasal extranodal NK/T-cell lymphoma among Brazilians, showed that EBV was more frequently found in adenoids than in palate tonsils, and that inflammatory lesions of the nasal and palatal regions did not present EBV (Altemani et al., 2002). The sequence of biopsies was performed to obtain a correct diagnosis, and this procedure is often necessary to confirm the neoplasm. The treatment remains confuse, and some authors believe that the best choice is to associate radiotherapy and chemotherapy (Takahashi et al., 2008), whereas other professionals have good results with chemotherapy only. Surgery is not an appropriate treatment without coadjuvant therapy. Most reported cases in literature seem to follow a similar course, including rhinorrhea, ulceration and necrosis of soft tissue, bone and cartilage of the face (such as the hard palate or nasal septum), leading to perforation. Secondary infections and cachexia commonly lead to death (Califano et al., 1998). These characteristics may help the early diagnosis when biopsies are inconclusive.

In the presented case, the initial treatment was chemotherapy followed by radiotherapy, with good results. The hard palate was destroyed, but healthy mucosa could be observed. Several teeth were lost because of the bone destruction. Unfortunately, an aggressive recidive with a rapid course lead to lung metastasis and patient’s death.

Conclusion

The extranodal NK/T-cell lymphoma is a rare lesion that demands professional knowledge for early diagnosis. Several biopsies may be required to confirm the diagnosis. It is important to evaluate clinical conditions for early treatment.

References

Illustrations

Illustration 1

Figure 1: Clinical aspect - 1. Necrotizing lesion on palate and nasal septum. 2. Anterior view showing anterior maxilla compromised.

Illustration 2

Figure 2 Presence of atypical lymphoid cells among acute inflammatory infiltrate (H&E: 100X, 400X).
Illustration 3

Figure 3: After treatment: areas of necrose, and healthy tissue in the posterior region.
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