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CLINICAL CASE

Laryngotracheal NK/T lymphoma: Clinical case

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KEYWORDS
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Abstract NK/T-cell lymphoma is a rare condition with an aggressive course and poor prognosis. Historically known as "lethal midline granuloma", it generally appears in a midfacial location. We describe the case of a 22-year-old Colombian woman with laryngotracheal affectation, presenting with hoarseness and hemoptysis. CT scan and MRI showed severe laryngeal and tracheal destruction. The biopsy showed a polymorphic, lymphoid cell infiltrate with angiocentric and angiodestructive pattern. The immunohistochemical study confirmed the immunophenotype of the NK/T-cells: CD2+, CD56+ and cytoplasmic CD3+. The in situ hybridization and flow cytometry findings were: EBER+, TIA-1+ and perforin+. The patient died from complications of her disease, before undergoing oncologic treatment.

PALABRAS CLAVE
Linfoma T/NK;
Laríngeo;
Traqueal;
Granuloma letal

Linfoma T/NK laringotraqueal: caso clínico

Resumen El linfoma T/NK es infrecuente, de curso agresivo y mal pronóstico. Históricamente llamado «granuloma letal de línea media», su localización habitual es el macizo centro-facial. Presentamos un caso de localización laringotraqueal, en mujer colombiana de 22 años. Refería disfonía y hemoptisis. Presentaba desestructuración y necrosis laringotraqueal. En el estudio anatomopatológico existía infiltrado polimorfo y linfoide de disposición angioinvasora y angiodestructiva. La inmunohistoquímica confirmó el fenotipo de los linfocitos T/NK: CD2, CD56 y CD3 citoplasmático positivos. Epstein-Barr positivo. TIA-1 y perforina positivas. La paciente falleció por complicaciones de la enfermedad, sin poder recibir tratamiento oncológico.

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Introduction

Nasal, T/NK cell lymphoma, is a non-Hodgkin, extranodal variety of the T-cell group. It is characterised by its aggressiveness and negative development, with very poor response to current therapies. It is rare in Europe and the United States, with a higher incidence in Asia and Latin America. Usually located in the central facial area, it can also appear in the skin, intestine, testis, and various areas of the upper aerodigestive tract.[1,2] Laryngeal and tracheal locations are uncommon. The non-specific initial symptoms and the intense necrosis of the tissues involved do not favour the diagnosis, which is only confirmed by immunohistochemical analysis. The differential diagnosis must consider conditions that cause destruction of the central facial area such as autoimmune diseases (Wegener's, Churg-Strauss), other lymphomas and cocaine consumption, as well as granulomatous infections (syphilis, tuberculosis, leishmaniasis, etc.) and other neoplasms.[3]

Clinical case

We present the case of a 22-year-old Colombian woman who was referred to us from another centre and in whom no aetiology was found after being admitted and studied for one month due to naso-laryngotracheal lesions. She complained of progressive dysphonia and haemoptysis of 6 weeks' evolution. She had a history of chronic rhinosinusitis without polyposis, treated with nasosinusal endoscopic surgery at age 19. Three years later, just before travelling to Spain, she presented a purulent nasal discharge, nasal blockage, epistaxis and nasal septal ulceration, which progressed to septal perforation and collapse of the nasal dorsum. It was apparently controlled with medical treatment.

On admission, she was eupneic, dysphonic and in good general condition. She presented septal and nasal crusting. Fibroscopy revealed severe laryngeal and tracheal destruction, in addition to inflammation and necrosis, bilateral laryngeal hypomobility and preserved glottic passage. The thoracic trachea and other lower respiratory tract structures were normal. No cervical lymphadenopathy was observed. Serological and autoimmune tests resulted negative. Renal function was normal.

Septal perforation and postoperative changes of medial bilateral antrostomy were revealed in CT and MRI scans, as well as signs of laryngeal mucosa ulceration and destruction of the first tracheal rings, with an absence of tracheal wall at this level. Paralaryngeal air was detected with an unidentified perforation site. No cervical lymphadenopathies were observed (Figure 1, Figure 2). No abnormalities were found in chest, abdomen and pelvis.

Biopsy: polymorphous inflammatory infiltrate, predominantly lymphocytic, of small and medium size and normal aspect, which infiltrated and destroyed the vessel walls. No granulomas or giant cells were found. Immunohistochemical analysis revealed phenotype of positive T/NK lymphocytes: cytoplasmic CD2, CD56 and CD3. In situ hybridisation and flow cytometry: intensely positive Epstein-Barr virus and positive for perforin and TIA1.

With these results, we reached the final diagnosis of extranodal T/NK cell lymphoma, nasal type. The patient suffered cardiac arrest from upper airway bleeding, post-anoxic encephalopathy and death.

Figure 1 Cervical CT scan: laryngotracheal destruction; paralaryngeal gas (arrow).

Figure 2 Cervical CT scan. Absence of tracheal rings; tracheal lumen in contact with left carotid sheath (black arrow), thyroid gland and oesophagus. Pre-thyroid gas (white arrow).

Discussion

Our attention was drawn to the stabilisation of the facial destructive symptoms, to the extent that hoarseness and haemoptysis were the main reasons for consultation, but we were not able to find an explanation for them. On physical examination, the nasal lesions were apparently "inactive".

We initially considered an infectious disorder due to the inflammation and necrosis, but this was ruled out by negative

biopsy, serology and cultures. The urine test for inhaled drugs was negative. We considered Wegener's disease in the diagnosis, due to the nasal and laryngotracheal lesions, but the renal function was normal and autoimmunity tests (including c-ANCAs) were negative; nevertheless, there still remained the possibility of a localised form of Wegener's disease, which represents 20% of cases, is more common in women, results in negative c-ANCA studies and presents normal renal function.[4]

Due to the inconsistency of this diagnosis and the central facial involvement, which characterises T/NK lymphoma (historically called "lethal midline granuloma"), we turned our attention to conventional histology. This revealed a non-specific angioinvasive and angiodestructive infiltration that may characterise T/NK lymphoma. Through the immunohistochemical analysis, we confirmed the phenotype of T/NK lymphocytes and, consequently, the diagnosis.

In this case report, the general condition of the patient was good and the analyses revealed no diagnostic data. This lack of correlation between the extent of the injuries and changes in the analysis is typical of T/NK lymphoma.[3] Unfortunately, it was not possible to initiate cancer treatment due to an episode of cardiac arrest and subsequent death. Although the location of the lesions was unusual, the aggressiveness was a feature consistent with this disease.

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