

CASE STUDY

Primary manifestation of Hodgkin lymphoma in adenoid. About a case

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Received May 10, 2009; accepted October 30, 2009

KEYWORDS

Hodgkin's disease;
Waldeyer's ring;
Extranodal

PALABRAS CLAVE

Enfermedad
de Hodgkin;
Anillo de Waldeyer;
Extranodal

Abstract

Lymphomas are the second leading cause of malignancy in head and neck. Hodgkin's disease (HD) accounts for only 10%–35% of all cases, where the lymph node is affected in 70%–80%. We present the case of a patient with HD with extranodal involvement, given the rarity of this entity.

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Manifestación primaria de linfoma de Hodgkin en adenoides. A propósito de un caso

Resumen

Los linfomas constituyen la segunda causa de neoplasia en cabeza y cuello. El linfoma de Hodgkin representa solo el 10%–35% de todos estos casos, donde la afectación ganglionar está presente en el 70%–80%. Presentamos el caso de un paciente con linfoma de Hodgkin de afectación extranodal, dada la rareza de esta entidad.

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Case study

We present a 39-year-old male with a history of tonsillectomy in childhood, who referred no toxic habits and with a maternal grandmother who had suffered chronic lymphocytic leukaemia.

The patient was seen in consultation due to nasal snoring and respiratory failure associated with daytime fatigue, more pronounced during the previous year and with no other associated symptoms. He was a professional driver who worked shifts. An inter-consultation with the Sleep Disordered Breathing unit was requested, which resulted in a diagnosis of mild sleep disorder.

An anterior rhinoscopy exploration showed a right septal deviation with compensatory turbinate hypertrophy. Nasofibroscope enabled us to identify a nasopharyngeal mass, compatible with adenoid hypertrophy. Rhinomanometry showed a greater air

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Figure 1 MRI showing a cervical mass of 2x3 cm, with a primary craniocaudal axis, without contrast uptake.

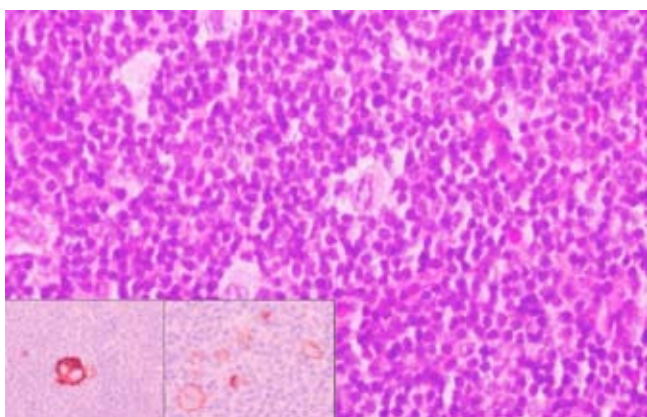


Figure 2 H-E: Reed-Sternberg cells with immunophenotypes CD15 (+) and CD30 (+), with lymphoid cell background.

resistance in the right nostril, without much improvement after vasoconstrictor treatment, with the rest of the ENT exploration being normal.

Given the worsening of the clinical manifestations in the previous year and the size of the nasopharyngeal mass, a biopsy, which revealed follicular lymphoid hyperplasia, and an MRI, which objectified a mass of 2.3 cm in diameter, consistent with adenoid tissue hypertrophy, were performed (Figure 1).

Considering the aforementioned clinical manifestations and the examination performed, the patient underwent septoplasty with turbinectomy and adenoidectomy. The anatomopathological report was classical Hodgkin's lymphoma (HL), rich in lymphocytes, with no significant presence of Epstein Barr virus (EBV) in the *in situ* hybridization study (Figure 2).

The patient was referred to the Haematology Service, eliminating any other location and establishing the diagnosis of stage IE-A classical HL rich in lymphocytes. After treatment with 2 cycles of chemotherapy (ABDV) and 20 Gy locoregional radiotherapy, the patient presented a complete remission of the condition.

Discussion

Non-Hodgkin lymphomas are the most common cause of head and neck lymphoma, with HL being responsible for only 10%-35% of cases. The classical HL manifestation is nodal, present in up to 70%-80% of cases, with extranodal manifestation being rare. The primary involvement of Waldeyer's ring in HL has an incidence of 1%-5%.¹ The patient described here presented a case of HL located in the adenoids, which did not affect other regions and did not have associated lymphadenopathy.

In published series of HL with primary involvement of Waldeyer's ring, the average age at onset is 40-50 years, with a higher incidence in elderly patients, in contrast to nodal HL, which has a bimodal distribution with a first peak between 25-30 years and a second at 45 years.² The incidence in males is 1-3 times higher than in females.¹

Given the rarity of this disease, its diagnosis should be confirmed by immunohistochemistry, with detection of Reed-Sternberg cells with CD15 (+), CD30 (+), CD45 (-), CD20 (-) and EMA (-) immunophenotype, enabling a differential diagnosis with Hodgkin lymphoma to be established.³ In most published studies, mixed cellularity is the most common histological subtype in extranodal HL, while the nodular sclerosis subtype is predominant in nodal affection.⁴ Association with EBV has been described in up to 40% of patients with HL, with the mixed cellularity subtype being the most heavily implicated.⁵ In the case presented, the results of immunohistochemical studies ruled out non-Hodgkin's disease, showing positivity for CD15 and CD30. However, the subtype corresponded to classical Hodgkin's, rich in lymphocytes; *in situ* hybridization studies did not detect the presence of EBV. It should still be noted that the sensitivity of this test is one copy for each one (2 cells), with a negative result not excluding the presence of EBV.

Advanced stages of rhinopharyngeal HL are rarely seen. In the review of 7 rhinopharyngeal HL carried out by Anselmo,⁶ all cases presented early stages (I-II) and all showed complete remission after chemotherapy (2-4 cycles) associated with radiotherapy (25-30 Gy), with an average monitoring of 72 months. The case presented here is consistent with the

results described, although monitoring was not more than 8 months.

Conclusion

Extranodal involvement of HL has a low incidence, of 1%5%

Diagnosis is usually carried out in early stages, being more common in males.

Immunohistochemical studies are needed to establish a differential diagnosis with non-Hodgkin lymphoma, which is the most common in head and neck.

Treatment is chemotherapy and radiotherapy, associated with a good prognosis.

Conflict of interest

The authors declare no conflict of interest.

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