CASE STUDY

Anaplastic Large T-Cell Lymphoma of the External Ear in Childhood

Verónica Rodriguez, a,* José María Perolada, a Isabel Ibañez, a Amets Fernández b

a Servicio de Otorrinolaringología, Hospital Universitario y Politécnico La Fe, Valencia, Spain
b Servicio de Anatomía Patológica, Hospital Universitario y Politécnico La Fe, Valencia, Spain

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Abstract Anaplastic large T-cell lymphoma is a very rare disease in childhood. The most common locations are lymph nodes and skin, while the external ear location is uncommon. We present the case of a 6-year-old child with an earlobe tumour. Surgical treatment was performed and the anatomicopathological results showed anaplastic large cell lymphoma. Radiological tests were negative and there was no systemic involvement.

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Linfoma T anaplásico de células grandes de pabellón auricular en la infancia

Resumen El linfoma anaplásico de células grandes es una patología rara en la infancia. La localización más frecuente es en la piel y nódulos linfáticos, siendo infrecuente la afectación del pabellón auricular. Presentamos el caso de un niño de 6 años con tumoración en el lóbulo de la oreja. Se realizó tratamiento quirúrgico y el resultado de la anatomía patológica mostró linfoma anaplásico de células grandes. Los estudios radiológicos fueron negativos y no hubo afectación sistémica.

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Introduction

Primary cutaneous lymphoma is a neoplastic proliferation of cutaneous B or T cells, with no evidence of systemic disease at the time of the diagnosis or for a period over 6 months after full clinical evaluation.1–8

According to the classification proposed by the World Health Organisation and the European Organisation for Research and Treatment of Cancer (WHO/EORTC, 2008), the primary cutaneous lymphoproliferative disorders of CD30-positive T cells include anaplastic large T cell lymphoma (ALCL), lymphomatoid papulosis and limit cases.3–5

ALCL accounts for 25%–30% of cutaneous lymphomas and is the second most frequent group. It mainly affects adults

* Corresponding author.
E-mail address: pecasve@hotmail.com (V. Rodriguez).

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aged between 40 and 60 years and is predominant among males, being rare in the paediatric population.4-11

The lesions are most frequently located in the limbs. They usually appear as individual nodules or tumours, although they can also do so as papules with or without ulcerations. They may also be multifocal in 20% of cases.1-10

Extracutaneous involvement may occur in 10% of cases, and consists in nodal involvement carrying a high rate of recurrence.3,4,6,7

The diagnosis is obtained through histological study and is characterised by a dense nodular infiltrate of tumoural, CD30+ large cells affecting the dermis and extending into the hypodermis. The cells show an activated T phenotype (CD2+, CD3+, CD4+, CD45RO+, CD25+, CD30+). They may express ALK (Anaplastic large cell Lymphoma Kinase protein) or not, although expression is related to the systemic form, whose prognosis and treatment are very different.3-7,12 The recommendation is to conduct disease extension studies through thoracic and abdominal computed tomography (CT) scans, bone marrow aspiration, cerebrospinal fluid cytology and bone scintigraphy scans.1-10

Cumulative survival at 10 years is over 90% in patients with primary cutaneous involvement.1-9

Treatment options vary depending on the degree of cutaneous or systemic involvement, and range from strict monitoring to systemic chemotherapy in severe cases.3-7

We report the case of a male child with primary, cutaneous, anaplastic, large cell lymphoma located in the earlobe.

Case Report

We present the case of a 6-year-old boy referred to our hospital due to a lesion with a torpid evolution in the right earlobe, with an infectious aspect, with local inflammatory signs and a hard consistency. The injury was drained through a puncture at another centre, without any purulent material being obtained. There was no response to oral antibiotics.

Upon admission, we established empirical intravenous treatment with cefepime at 1g/8h and teicoplanin at 200mg/12h. The lesion continued to increase in size so, given its accessibility and reduced dimensions, we decided to avoid cytology with fine needle puncture-aspiration and instead carry out its surgical excision in order to perform a diagnostic and microbiological study of the specimen. The result showed an absence of infection and the presence of a diffuse infiltrate of loose pleomorphic cells with large nuclei and small lymphocyte infiltrate, with hyperchromatic nuclei and with eosinophils and macrophages (Fig. 1). These cells were lymphoid, negative for ALK and expressed CD30, CD3, CD5, CD20, CD43 and CD79 infiltration. This data pointed to the diagnosis of anaplastic CD 30+/ALK− T cell lymphoma.

We conducted an extension study through cervicothoracic and abdominal CT scans, bone marrow aspiration, cerebrospinal fluid study, cervical ultrasound and bone scintigraphy scan with technetium-99 (Fig. 2). The test results were negative for lymphoma infiltration. Upon recommendation from paediatric oncology, since the first intervention took place for diagnostic purposes, we decided to extend the surgical margins, which were negative for disease. The resulting defect could be closed without the use of flaps. In these conditions, we opted for strict monitoring without establishing any further treatment. After 12 months follow up, the patient is currently asymptomatic.

Discussion

ALCL is a rare condition which was first described in 1985 by Stein et al.8 as a pleomorphic cell malignancy with expression of the CD30+ marker, which, until that moment, was considered unique to Hodgkin’s disease. Subsequently, Mann et al.9 and McCluggage et al.10 described 2 morphological variants with predominance of neutrophils and eosinophils, respectively.

In 2008, a new classification was proposed by the WHO/EORTC which unified the clinical concepts and differentiated cutaneous lymphomas into 3 types: T-NK cell lymphomas, cutaneous B cell lymphomas and haematological precursor neoplasms.3

Over the past 8 years there have been different reports of series with cases of anaplastic lymphomas with cutaneous involvement in patients aged between 12 and 80 years. Its occurrence in childhood is very rare. The most common location is in the face and limbs.6-13

Figure 1 Pathological anatomy: haematoxylin-eosin staining (20x), with diffuse infiltrate of isolated pleomorphic cells with large nuclei and small lymphocytic infiltrate.

Figure 2 Bone scintigraphy scan with technetium-99 showing no evidence of systemic involvement.
There have been documented cases associated with Epstein–Barr virus, trauma, immunosuppressive states such as those caused by heart or kidney transplant and infection with HIV.5–13

According to various published data, our case appeared at a highly unusual age (6 years) and was located in an area not previously documented in the literature (earlobe).

Diagnosis of the disease is difficult to establish, especially in paediatric patients, as it is necessary to bear in mind when faced with a seemingly infectious process with an unfavourable evolution.1,3,5 A thorough histopathological analysis, including immunohistochemical markers such as CD30 and ALK, is important to establish the diagnosis, since the prognosis and management will differ depending on the results.3–5,8

The differential diagnosis must include other, more common entities which cause a diffuse inflammatory infiltrate, such as insect bites, hidradenitis suppurativa, stasis ulcer, ruptured cysts, bacterial, viral and fungal infections.5,6,11–15

The form of presentation as a rapidly growing tumour is very alarming. Thus, in the past, it led to very aggressive chemotherapy being administered, which is not recommended at present, as it does not prevent cutaneous recurrences.6,8–10,12 Currently, chemotherapy is only administered in cases with extracutaneous extension (appearing in 10% of cases). ALK marker negativity also justifies a conservative management, since its positivity has been associated with disseminated cases.3–10

In our case, the patient developed satisfactorily 12 months after excision of the tumour. Nevertheless, all cases require long-term monitoring in order to diagnose cutaneous recurrences.

Conflict of Interests

The authors have no conflict of interests to declare.

References