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Case Report
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Cutaneous Nodules Revealing Large Cell Anaplastic Lymphoma ALK+

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Abstract

Anaplastic Large Cell Lymphoma (ALCL) encompasses at least 3 clinically distinct entities. The entity ALK with systemic ALCL is rare. In the infant population skin involvement is rarely revealing. Diagnosis and early management improves prognosis. We report an original case of a 7-year-old child in whom the skin involvement was indicative of neoplasia.

Keywords: Cutaneous nodules-large cell anaplastique lymphoma ALK+ -chemotherapy

Introduction

Anaplastic large cell lymphoma (ALCL) is an uncommon T-cell lymphoma defined according to the Revised European American Lymphoma and World Health Organization classifications as a distinct clinicopathological entity [1]. Cutaneous ALCL presents either as primary cutaneous disease or as secondary skin involvement due to the systemic disease. We describe an unusual systemic ALCL revealed by cutaneous nodules.

Case Presentation

A 7-year-old child with a history of herpetic encephalitis at 2 months of age, consult the emergency room for nodal lesions that have been evolving for 3 weeks with anaphylaxis. The examination of the skin revealed numerous nodular swellings which were erythematous-violin, not painful and slightly pruriginous. Some of which had a surface ulcerated, and others were covered by blackened crusts, sitting in the lower limbs, scalp and in left orbital (Figure 1). Adding to this, he had some erythematous and finely scaly macules in the trunk and back. The examination of the ganglionic areas found multiple cervical, spinal and inguinal nodes (Figure 1), the largest of which measured 3cm of major axis sitting at the right inguinal level, fixed with erythematous opposite surface.

The biological examinations revealed a normochromic normocytic argenerative anemia at 5g/dl, without leucopenia or thrombocytopenia with. Renal and hepatic imaging was not abnormal. The cutaneous biopsy with immunohistochemical study confirmed the diagnosis of a cutaneous localization of malignant non-Hodgkin lymphoma with large ALK + anaplastic cells (Figure 2). The osteomedullary biopsy revealed a medullary invasion of a malignant tumor process with little differentiation. The extension report showed only tumor-like gonglions. The evolution was marked by a subsidence of the nodules after the 3rd cure of chemotherapy, CHOP protocol. Unfortunately the child died at the 6th cure by a sepsis.

Discussion

Anaplastic Large Cell Lymphoma (ALCL) was first described in 1985 based on the large pleomorphic cells expressing CD30, its propensity to invade sinusoids, and the cohesive appearance of the tumor [2]. ALCL encompasses at least 3 clinically distinct entities (primary cutaneous ALCL (pcALCL), anaplastic lymphoma kinase (ALK) with systemic ALCL (sALCL), and ALK without sALCL [3,4]. sALCL is usually positive for ALK and epithelial membrane antigen (EMA) expression; which are negative in pcALCL [3]. Our patient had CD30+, ALK+ and EMA+ with medullary invasion by tumoral cells. sALCL are known to be in advanced-stage disease at presentation. It primarily involves lymphnodes, although extranodal sites maybe involved [5]. In children, 18–25% of ALCLs develop skin manifestations during the course of the disease [6]. These manifestations rarely reveal the disease. Our patient had already developed cutaneous nodules 3 weeks previously.

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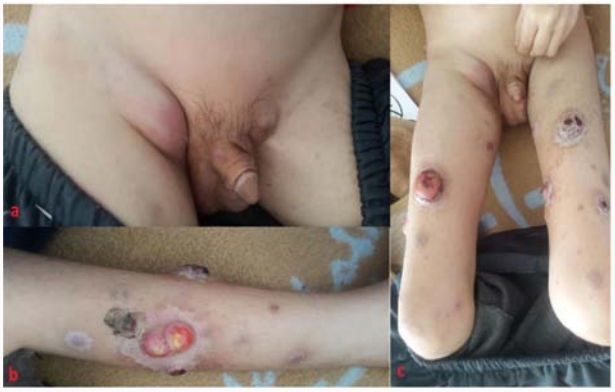

Figure 1 : A: clinical aspect of bilateral inguinal adenopathies. B and C: multiple cutaneous nodules of the lower limbs.

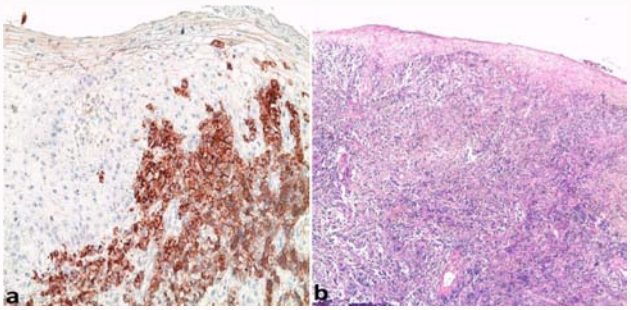

Figure 2: A: HES staining G x 50 -> Dense lymphoid dermal infiltration. B: IHC G x 200 -> CD30 (+).

The histopathology study allowed making the diagnosis. In children, the systemic form is more common and prognosis is worst when the skin is also affected [7,8]. sALCL are treated with chemotherapy containing antrhracycline such as CHOP [9]. Autologous and allogeneic stem cell transplantation may be beneficial in case of recurrence [9]. Brentuximab vedotin is an anti-CD30 monoclonal antibody targeting malignant CD30-positive cells, which shows also its efficiency in the treatment of refractory and recurrent systemic ALCL [10]. The decision was to treat our patient with CHOP protocol with a good improvement from the 3rd cure of chemotherapy.

Conclusion

We report an original pediatric case of sALCL AKL+. The cutaneous biopsy plays an important role in making this diagnosis. Early detection and treatment may provide a better outcome.

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