A rare pediatric case of cutaneous gamma/delta T-cell lymphoma

Un cas pédiatrique rare de lymphome T cutané gamma/delta

Abstract. Cutaneous γ/δ T-cell lymphoma (CGD-TCL) is a recent entity described in the newly revised World Health Organization-European Organization for Research and Treatment of Cancer classification of cutaneous lymphomas. Only a few cases have been reported, of which two pediatric cases. A 15 years old child with a 6 months history of polyadenopathy, cutaneous lesions, general edema and deterioration of general condition was hospitalized. Results from laboratory testing, cutaneous histopathology and immunohistochemistry showed a primary CGD-TCL. Staging was completed by a total body computed tomography. Therapy was planned with SMILE protocol. It is a highly aggressive tumor resistant to chemotherapy, immunotherapy, and radiation therapy. The GDTCL is characterized by a worse prognosis with a median survival of 15 months. Early diagnosis is essential and aggressive therapy is necessary.

Key words: cutaneous γ/δ T-cell lymphoma, WHO-EORTC classification, primary cutaneous lymphoma, SMILE, paediatric

Résumé. Le lymphome T cutané γ/δ (LTCGD) est une entité récemment décrite dans la Classification actualisée des lymphomes cutanés réalisée par l’Organisation mondiale de la santé et de l’Organisation européenne pour la recherche et le traitement contre le cancer. Seuls quelques cas ont été rapportés, parmi eux 2 cas pédiatriques. Un enfant de 15 ans a été hospitalisé pour bilan d’une polyadénopathie avec lésions cutanées, œdème et altération de l’état général évoluant depuis 6 mois. Le bilan biologique, l’étude anatomopathologique et immunohistochimique des lésions a révélé un LTCGD. Un scanner a complété le bilan d’extension. Un traitement par le protocole Smile a été décidé. Il s’agit d’une pathologie extrêmement agressive, résistante à la chimiothérapie, l’immunothérapie et la radiothérapie. Le pronostic est très mauvais avec une médiane de survie à 15 mois. Un diagnostic précoce et une prise en charge thérapeutique agressive sont indispensables.

Mots clés : lymphome T cutané γ/δ, classification WHO-EORTC, lymphome cutané primitif, Smile, pédiatrie

Primary cutaneous gamma-delta T-cell lymphoma (CGD-TCL) is a very uncommon and extremely aggressive tumor included since 2008 in the WHO classification as a rare sub-type of primary cutaneous peripheral T-cell lymphoma [1, 2]. About 40 cases have been described in the literature [1] with 42 years average age at onset of disease. We find only 2 cases about children except ours. We report the case of a 15 years old child with hepatosplenic involvement.

Case report

A 15 year-old patient without any pathologic history presented at our department with a 6 months history of polyadenopathy, cutaneous lesions, general edema and deterioration of general condition. Physical examination showed that he was 150 cm tall and weighed 45 kg. Performance status was 0. On his leg skin were showed localized (figure 1), dusky red colored plaques with peripheral erythematous patch (figure 2). The
Current practice

Figure 1. Multiple red colored lesions in primary cutaneous gamma-delta T-cell lymphoma.

Figure 2. Erythematous plaques of the leg in primary cutaneous gamma-delta T-cell lymphoma.

abdominal examination revealed hepatosplenomegaly. In the axillary regions on both sides there were indolent and moveable lymph nodes, in the neck were jugular, carotid and spinal lymph nodes measuring 1 to 1.6 cm. The ORL examination showed Waldeyer’s ring involvement.

Results from laboratory testing showed normochromics normocytic anemia with hemoglobin 8.6 g/dL on blood count. On serum tests, CRP was 14 mg/L, LDH: 535 UI/L, β2microglobulin 12.8 mg/L and hepatic tests were disturbed; ASAT:181 UI/L, ALAT: 112 UI/L, PAL: 769 UI/L, γGT: 362 UI/L.

Histopathology of skin lesions revealed a dense dermal and subcutaneous infiltrate of small to medium sized pleomorphic lymphocytes with numerous mitosis and epidermotropism. Immunohistochemistry showed positivity of CD3, CD4, CD56 and granzyme B, but was negative for CD 20 and CD30; a small number of cells was positive for CD8 and EBV.

In sum, according to the WHO-EORTC classification, the results were consistent with γδ T-cell lymphoma. Molecular pathology and PCR amplification for the γ chain of the T-cell receptor were not achieved. Computed tomography neck/thorax/abdomen/pelvis showed hepato-splenomegaly, ascite, bilateral jugular and carotid lymph nodes.

Therapy was planified with SMILE protocol (Japanese group) but the patient died first day of therapy.

Discussion

About 40 cases [3] of primary CGD-TCL have been reported worldwide. It is a highly aggressive tumor resistant to chemotherapy, immunotherapy [3], and radiation therapy. Since 2008 it has been included in the WHO classification for myeloid and lymphatic neoplasms as a rare subtype of primary cutaneous peripheral T-cell lymphoma [4]. Histologically, the δγ subtype can infiltrate from the subcutaneous fat to the epidermis [5]. Cytological studies show infiltrate composed of variably pleomorphic T-cell from medium-sized to large atypical lymphocytes with coarse chromatin; occasionally we find large blast cells with vesicular nuclei and prominent nucleoli. Epidermotropism is prominent in some cases like ours; apoptosis and necrosis are frequently seen as well as angio-invasion and vascular destruction [6].

Immunophenotyping is critical for diagnosis. The expression of CD2, CD3 and CD7 is variable, CD5 is commonly absent. CD4 and CD8 are usually negative although CD8 expression has been documented in rare cases [7]. Our patient cells express CD4 and rare CD8. CD56 and cytotoxic proteins like granzyme B are frequently positive. Our patient’s condition seemed to be related to EBV but the role of EBV in γδT-cell lymphoma is still discussed [7, 8].

Clinical features are variable: cutaneous patches, plaques, nodules or necrotic tumors are often ulcerated and primarily affect the extremities.

Patients are adults, average age at onset of disease is 42 years with an equal distribution of males and females. 3 children cases, including ours, were described to our knowledge [9, 10].

The GDTCCL is characterized by a worse prognosis due to frequent association with hematophagocytic syndrome (50%) [2], subcutaneous involvement [7], CD56+ [11], resistance to chemotherapeutic regimens, radiation therapy and immunotherapy. These elements probably explain the poor outcome in this group with a median survival of 15 months [12].

Early diagnosis is essential and aggressive therapy is necessary to help these patients; allogenic stem cell transplantation appears to be a promising future option [13] with
few cases reported. Other treatment with retinoid and narrow band UVB [14] is challenging and will require further study.

**Conclusion**

Cutaneous γ/δ T-cell lymphoma is a recent entity described in the newly revised World health organization-European organization for research and treatment of cancer classification of cutaneous lymphomas, and is characterized by the γ/δ T-cell receptor expression on atypical lymphocytes. Only a few cases of pediatric primary CGD-TCL have been reported, with an extremely aggressive course.

**Conflicts of interest:** none.

**References**


