RARE PEDIATRIC CUTANEOUS T-CELL LYMPHOMA

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RARE PEDIATRIC CUTANEOUS T-CELL LYMPHOMA (Abstract): We describe a rare case of an adolescent with T-cell cutaneous lymphoma, having a history of intense pruriginous, erythematous and hyperpigmented skin lesions. Several considerations regarding diagnostic work-up and treatment must be emphasized. Erythematous, pruritic patches, unresponsive to common topical/general treatment should raise the suspicion of more serious immuno-allergic or neoplastic conditions, among which, cutaneous lymphoma could be a rare, but a realistic possibility in pediatric field. Long term follow-up by a pediatric haemato-oncologist and dermatologist team is mandatory for these patients. **Keywords:** CUTANEOUS LYMPHOMA, CHILD, PARAPSORIASIS.

In pediatric oncology Non-Hodgkin's Lymphoma is the third most common solid tumor, and constitute 5-7% of all malignancies, most often located in abdominal and thoracic regions. However, Non-Hodgkin lymphomas may have atypical primary presentations like otorhinolaryngology (1), central nervous system (CNS) or skin. Cutaneous T-cell lymphoma (CTCL) is a non-Hodgkin lymphoma that commonly occurs in elderly individuals (2,3,4,5). It is characterized by skin infiltration of malignant T lymphocytes. Mycosis fungoides (MF) is a subtype of CTCL while Sézary syndrome is its leukemic variant. The incidence varies among different age groups, with only 0.5 - 5% of cases occurring before 20 years old (3). The wide variety of clinical features and its similarities with other dermatoses may delay the diagnosis in early stages of MF. An indolent course is noticed in children and adolescents diagnosed with MF restricted to skin involvement (5). These patients require a multidisciplinary approach-pediatric haemato-oncologists, pathologists and dermatologists to improve their outcome.

CASE REPORT

We report the case of an adolescent girl who presented with a 5-months history of generalized body patches. The lesions were highly pruriginous, with mixed, erythematous and hyperpigmented regions of different sizes and shapes, with an initial chest and abdomen concentration, and a subsequent diffusion towards the upper and low-

er limbs. A cutaneous tumor of 5 cm below the right costal edge noticed. (fig. 1). Associated inguinal lymphadenopathy was observed. The patient denied drug abuse and no history of exposure to chemicals or radiation could be found.

No hepatosplenomegaly, intraoral lesions or arthralgia were found.

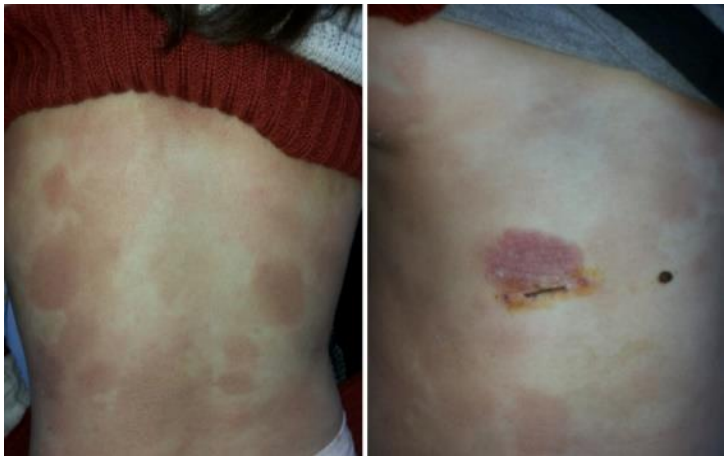

Fig. 1. Erythematous patches on the trunk and abdomen, erythematous plaque on the abdomen

Laboratory tests showed peripheral blood eosinophilia, with no other abnormalities. Antibodies against Cytomegalovirus, Epstein Barr virus, Toxoplasma were negative. Screening for B, C hepatitis B and C viruses, HIV, EBV, and CMV were performed for differential diagnosis of skin lesions and confirmation of chronic hepatitis and hepatic neoplasms (4).

Biopsy samples from skin lesions, lymph nodes and cutaneous tumor and subsequent histopathological and immunohistochemical examination were performed. Both skin and tumor biopsy revealed the presence of atypical epidermotropic lymphoid infiltrate with cerebriform nuclei extend into the dermis, suggestive for Mycosis fungoides (fig. 2). Also, small plaque parapsoriasis was present in the skin lesions. Lymph node biopsy revealed reactive lymphadenitis. The biopsy samples were sent to Timisoara County Emergency Clinical Hospital for further investigations. The immunohistochemistry analysis revealed positive CD2, CD3, CD4, CD5, CD7, CD8 markers and negative CD20, CD30 markers in the atypical lymphocytes from skin and tumor samples, confirming the diagnosis of MF (fig. 3).

Computed tomography of the neck, chest, abdomen and pelvis showed multiple enlarged lymph nodes in the cervical chain, axillary region (measuring up to 2.62 cm), mesentery (measuring up to 1.05 cm) and in the inguinal region (measuring up to 1.82 cm).

The bone marrow aspiration showed normal cellularity at different stages of maturation. *Clinical, laboratory and histopathological* findings established the diagnosis of Mycosis Fungoides, T3N0M0B0 - tumor stage, IIB, according to International Society for Cutaneous Lymphomas/European Organization for Research and Treatment of Cancer (ISCL/EORTC) revision to the staging of MF and Sezary Syndrome (8).

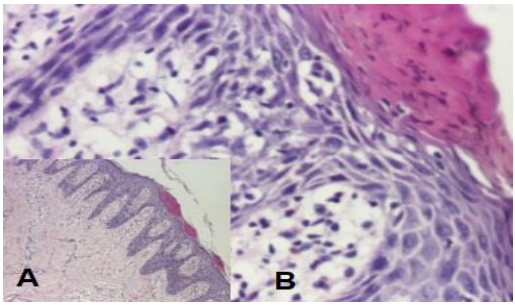

Fig. 2. A) Infiltrate into the epidermis and dermis (^{x}40 HE stains);
B) Infiltrate with abnormal cells with cerebriform nuclei, epidermotropic infiltrations of malignant cells (^{x}200 HE stains).

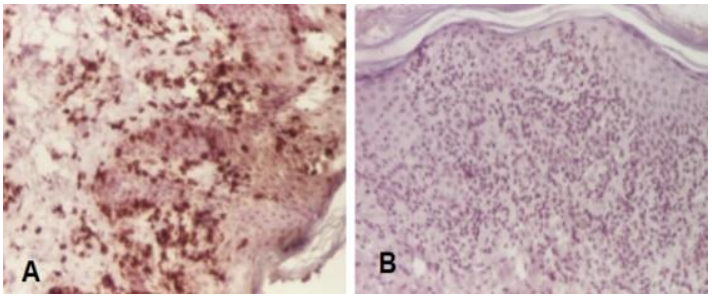

Fig. 3. A) Immunostaining - Abnormal cells positive for CD3(^{x}100);
B) Immunostaining - Abnormal cells negative for CD20 (^{x}100)

An initial regimen treatment with moderate potency topical steroids concomitant with narrowband ultraviolet B (NB-UVB) were used, followed-up by chemotherapy - cyclophosphamide, doxorubicin, vincristine and prednisone (regimen CHOP)-12 cycles, given in 21 days interval, and narrowband ultraviolet B-10 phototherapy sessions. CHOP chemotherapy sequence consisted in: Prednisone 100 mg PO daily Days 1 to 5, Doxorubicin 50 mg /m² IV Day 1, Vincristine 1.4 mg /m² IV (max 2 mg) Day 1, Cyclophosphamide 750 mg /m² IV Day 1.

The computed tomography and PET-CT were performed at the end of the treatment and showed no abnormalities. We noted a partial clinical response with the disappearance of pruritus and improvement of the skin lesions.

DISCUSSION

According to our previous experience (9, 10). pediatric neoplastic diseases may include some uncommon type of malignancies, requiring different confirmation procedures within the diagnosis process.

CTCL are quite rare among children with MF being a T-cell lymphoproliferative disorder and the major subtype of CTCL (2, 3). Data on clinical features, management, treatment response, disease progression in children are limited. MF is difficult to be distinguished from other entities in early stages due to its clinical polymorphism, mimicking benign skin disorders, that frequently occurs in pediatric age, such as eczema, atopic dermatitis, pityriasis, psoriasis. Therefore, consequently multiple biopsies may be necessary for

an accurate diagnosis (11, 12, 13).

In our case, the diagnosis of MF was suggested by histological and immunostaining parameters. Also, the association with small plaque parapsoriasis was observed in our patient.

Parapsoriasis manifests as large plaque and small plaque parapsoriasis (SPP). Both are caused by T-cell infiltration in the skin. If large plaque parapsoriasis is the earliest stage of CTCL (14,15), SPP is a benign form (16), but there were reported cases of progression to MF in adult population (17).

Our patient was diagnosed in an advanced stage-tumor stage IIB according to International Society for Cutaneous Lymphomas/European Organization for Research and Treatment of Cancer revision to the staging of MF and Sezary Syndrome (8).

Topical and systemic corticosteroids are used for treating advanced stages of MF. Systemic chemotherapy along with phototherapy is the most useful treatment schemes for advanced stages (4,18,19). In our case, no hypertension occurred, although it is known that this treatment regimen is prone to such complication (20, 21, 22). One should consider early diagnosis, fast and effective communication with stakeholders, as well as reflection upon the moral values when life is at stake, to a good prognosis (23, 24, 25, 26). Despite the several therapies reported, none of them is well-defined for children and adolescents. Our patient completed twelve cycles of systemic chemotherapy and narrowband ultraviolet B. Currently she is treated only with topical corticosteroids. Follow-up period for our patient is 2 years. She didn't develop so far, any complication or disease progression, but still has an inferiority complex about her aesthetic look.

CONCLUSIONS

We reported a rare case of juvenile-onset of MF. Long follow-up period is needed for our patient. Further studies are required for a better understanding of the outcome and prognostic implications of MF in pediatric age.

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