Case Report

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Hypo pigmented Mycosis Fungoides in Pediatrics: Case Report

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Abstract

Mycosis Fungoides is a malignant neoplasm originating in T lymphocytes, which mainly involves the skin and can present systemic involvement involving the bone marrow, lymph nodes and various organs. It is the most common type of cutaneous T-cell lymphoma and may have clinical varieties, predominantly hypopigmented. Lymphomatoid papulosis can precede this neoplasm in 5-20% of patients. We present the case of a 10-year-old female schoolboy with lymphomatoid papulosis and that 5 years later presented Hypopigmented Mycosis Fungoides.

Keywords: Hypopigmentation; Mycosis Fungoides; Lympho matoid Papulosis; Lymphoma; T-Cell; Cutaneous.

Introduction

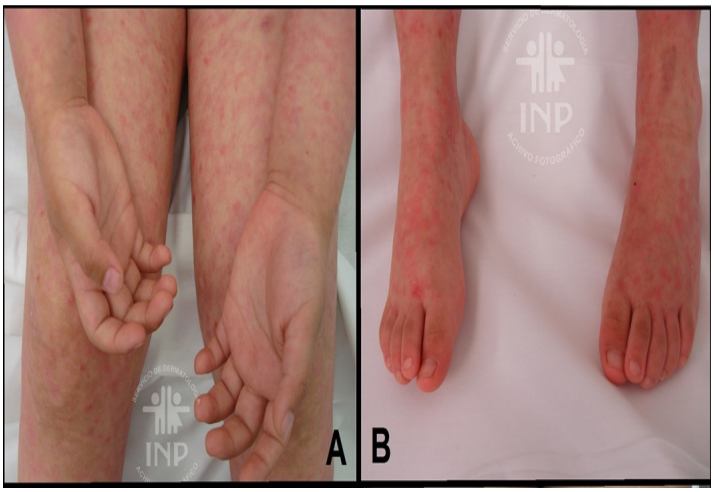
Mycosis Fungoides (MF) is a cutaneous epidermotropic T-cell lymphoma (CTCL), characterized by a clinical course indolented histologically by a proliferation of CD4 T lymphocytes, showing an affinity for the skin, and especially for the epidermis (epidermotropism) [1]. MF is the most common form of CTCL, accounting for 54-72% of cases; however, in childhood and adolescence it is unusual, assuming only 0.5%-5% of all cases. Hypochromic or hypopigmented MFH (MFH) is the clinical variant with the highest incidence in childhood [2]. There is a case of female schoolchildren who presented lymphomatoid papulosis and subsequent evolution to MFH.

Case Presentation

Female who begins suffering at 2 years of age with the presence of dermatosis characterized by plaques of different sizes, pink color, well-defined edges and presence of fine scale on the central surface, present mainly in the trunk and extremities, with the presence of intense pruritus, biopsy of the lesions is performed being compatible with chronic pityriasis liquinoid, so management with psolarenos and sun exposure is initiated, with apparent improvement and control of the disease.

At 5 years of age he presents reappearance of lesions characterized by papules with fine scale on the surface as well as hypochromic macules of poorly defined limits and nodules with central necrosis and presence of blood crust, which were disseminated to all body segments respecting face, palms and soles (See (Figure A and Figure B); according to the new clinical findings it is decided to perform another skin biopsy. The histopathological study reports lymphoproliferative disorder of T cells CD30, CD4, CD10, CD5, CD3, CD8, CD45, CD68 and KAPPA positive, compatible with Lymphomatoid Papulosis. During her evolution, the patient presents multiple exacerbations of the disease, meriting management with topical steroid during these episodes, achieving remission of the picture through management with psoralenos and topical steroid.

FigureA and B: Lymphomatoid Paupulosis: Lymphomatoid rash on extremities.



At 10 years of age it restarts with lesions characterized by hypopigmented macules of irregular and ill-defined limits, some with fine scale on its surface, and others of erythematous type, slightly infiltrated, which were predominantly in the abdomen and extremities, as well as non-photoexposed areas (See (Figure C and Figure D); before the change in the clinic of the lesions it is decided to perform a new biopsy, which was compatible with epidermotropic T lymphoma (hypopigmented Mycosis Fungoides), with Immunohistochemistry CD3, CD4, CD5, CD7, CD8, CD20, Granzima B negative and TIA 1 negative.

FigureC: Mycosis Fungoides: Reddish nodule with slight peeling.

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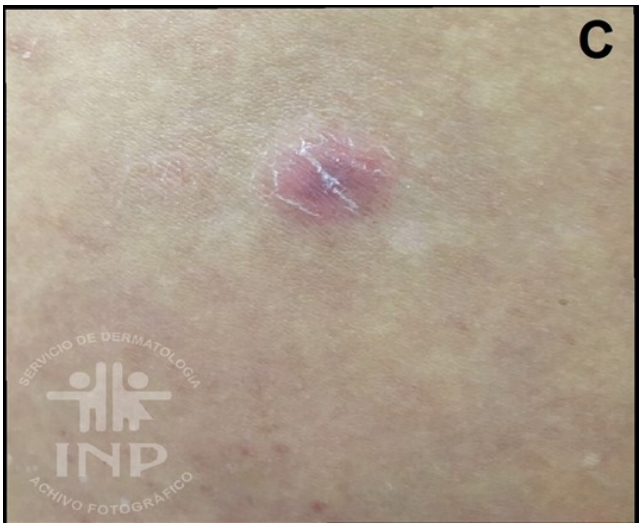
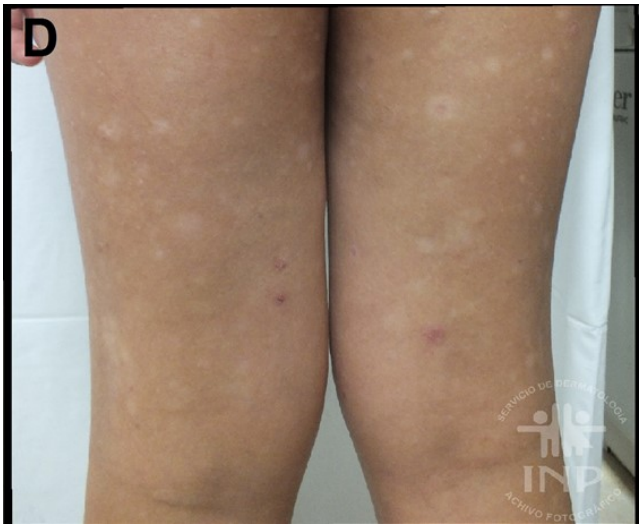


FigureD: Mycosis Fungoides Hypopigmented: Hypopigmented macules on thighs.



Given the new pathological diagnosis, extension studies are carried out to determine systemic condition, so PET/CT FDG is performed in which no malignant hypermetabolic activity was found.

Discussion

Primary cutaneous lymphomas (PCLs) belong to a heterogeneous group of malignant lymphoproliferative neoplasms, which mainly affects the skin without systemic involvement at the time of diagnosis (visceral, bone marrow or lymph nodes) [3]. Up to 5-20% of cases may be previously associated with lymphomatoid papulosis which is a T-cell lymphoproliferative disorder, CD30+, at risk of developing secondary lymphomas, a condition that is rare among adults and even rarer among children [4].

Several clinical variants of MF have been described: Hypopigmented, hyperpigmented, ichthyosiform, pityriasis lichenoid, granulomatous, folliculotropic, pagetoid reticulosis, purpuric, hyperkeratotic, and warty [5]. Purely , MFH is rare. The first case of MFH was described in 1973 [6]. The mean age of patients at diagnosis is between 40 and 60 years of age for classical MF, unlike hypopigmented MF that usually occurs at an early age, the average being 18 years of age. MF is more common in men, and is seen more frequently in African Americans relative to Caucasians [7].

MFH accounts for approximately 17 to 59% of childhood mycosis fungoides cases; however, the actual frequency of hypopigmented mycosis fungoides is unknown and could even be underestimated because it is often confused with some of its differential diagnoses, such as vitiligo, pityriasis alba, hypochromiant solar dermatitis, post-inflammatory hypopigmentation, etc., [2]. So it can represent a diagnostic challenge, as in the case presented in which the transformation from the lymphomatoid papulosis can make the approach even more difficult and makes it a diagnostic challenge, such as in the case presented in which the transformation from the lymphomatoid papulosis can make the approach even more difficult and makes it a diagnostic challenge. pathological and immunohistochemical confirmation is necessary to provide the definitive diagnosis.

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