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Case Report

Childhood Hypopigmented Mycosis Fungoides: The Greatest Imitator

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Abstract

Mycosis Fungoides (MF) is the most common type of cutaneous T-cell lymphomas, especially found in individuals over the age of 50. The incidence of childhood MF is low, only about 0.01-0.03% per year. The clinical manifestations of MF show great variation, with scaly erythematous patch as the most common presentation. In children, hypopigmented patch manifests frequently and can mimic several skin disorders, from leprosy to post inflammatory hypopigmentation. We reported a case of a 5-year-old girl with multiple red patches on her trunk and extremities. Our first differential diagnoses were leprosy, erythema annulare centrifugum, and granuloma annulare. Histopathology and immunochemistry findings correspond to hypopigmented MF. She was treated with mometasone furoate 0.1% cream and Narrow Band Ultraviolet B (NBUVB) for over a year.

Introduction

Mycosis Fungoides (MF) is the most common cutaneous T-cell lymphoma found in adult and elderly [1-5]. The incidence of MF is 0.29 cases per 100.000 inhabitants per year, with approximately only 0.01-0.03% seen in children every year [1,6]. The clinical spectrum of MF shows a wide variety, with scaly erythematous patches as the most frequent variant of MF found in children [2,5]. Hypopigmented MF is one of the rarest types of MF, but is found more commonly in children and adolescents [2-4,7]. Internal organs and lymph nodes involvement are very rare in children [2]. The diagnosis of MF is commonly delayed because the clinical and histopathological findings often imitate other benign skin diseases, such as eczema, vitiligo, pityriasis alba or pityriasis lichenoides [6,7].

We report this case because of its rare incidence,its difficulty in making the diagnosis as well as to treat. Early detection, the choice of effective therapy, and periodic evaluation may provide a good prognosis for hypopigmented MF in children [8].

Case History

A 5-year-old girl presented with erythematous patches that had started on her abdomen and spread to her back and extremities in 6 months. She was diagnosed as eczema and treated with topical steroid. Some patches slowly faded and became hypopigmented. She was then suspected to have leprosy. The patches were not numb, itchy, nor painful. On physical examination there were multiple erythematous patches and plaques, some with central hypopigmented (Figure 1). The sensibility test was difficult to be performed, and there was no peripheral nerves enlargement. The differential diagnoses were leprosy, atopic dermatitis, erythema annulare centrifugum, and granuloma annulare. A punch biopsy was done on the hypopigmented lesion. Histopathological findings showed a collection of mixed inflammatory cells consisting of lymphocytes and histiocytes that form epithelioid granulomas primarily around blood vessels and nerves, which resembled leprosy (Figure 2). Interestingly, we also found atypical cells with epidermotrophism that is suggestive of MF. Submitted stains of Ziehl Nielsen and Fite Faraco were negative. Immunohistochemical examination showed CD20-, CD3+, CD4+, CD8+, and CD56+. These results still could not exclude T-cell lymphoproliferative lesions or Natural Killer (NK)/T cells lymphoma, so we proceed with EBER in-situ hybridization examination, which showed negative results. Her peripheral blood examination within normal limits and no Sezary cell found. The immunophenotyping showed no blast cell (CD34+) from hematopoietic cell population (CD45+), which excluded acute leukemia. There was no involvement of peripheral lymph nodes and other internal organs. The patient was then diagnosed as hypopigmented MF. She was treated with Narrow Band Ultraviolet B (NBUVB) three times a week which then was tapered until once a week for over a year, with a total cumulative dose of 117.148 J/cm^2. She was also given mometasone furoate 0.1% cream once a day for 5 days in a week and tapered to twice a week. After 1 year of treatment, most of the patches were clear (Figure 3). However, she still had a hyperpigmented macule on her right arm, where then we performed a punch biopsy. Histopathological findings of the old hyperpigmented lesion showed some epitheliod granulomas, which was negative of mycobacterial organism, and the immunohistochemistry revealed negative for mycosis fungoides.

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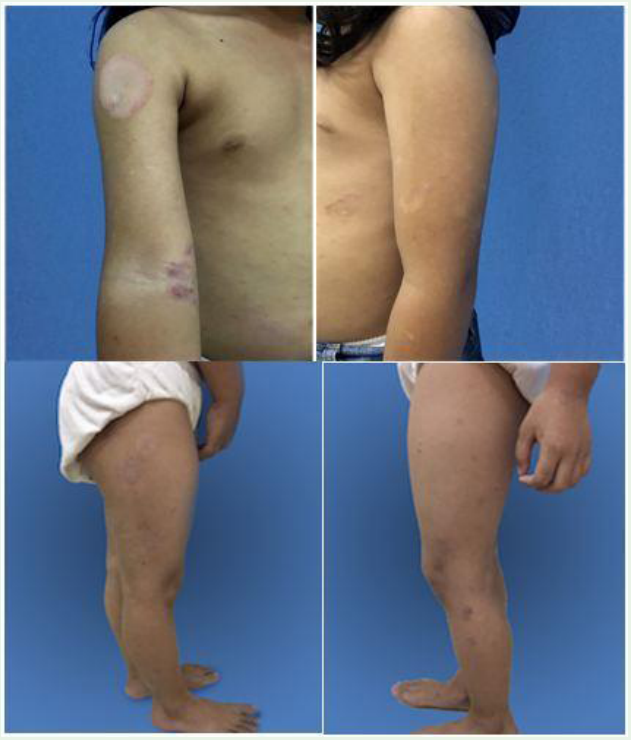

Figure 1: Baseline pictures.

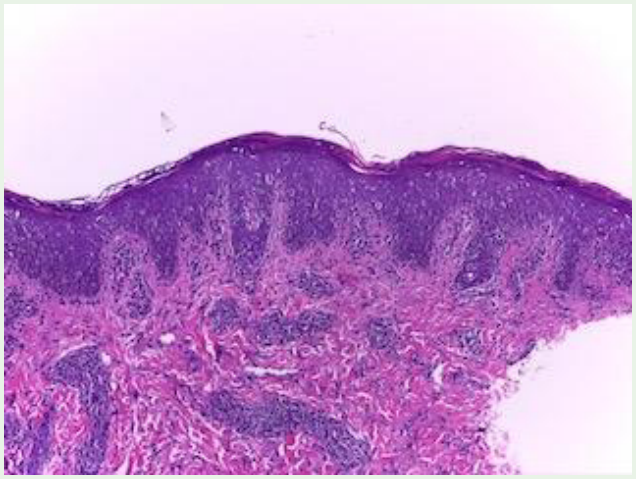

Figure 2: H & E, 4x Histopathology showed epithelioid granulomas primarily around blood vessels and nerves and epidermotrophism.

Discussion

Mycosis Fungoides (MF) is one of the most common cutaneous lymphomas [7,8]. Hypopigmented MF is an atypical variant of MF which is very rare and unique, mostly found in Asian and dark-skinned individuals [5,7]. Cutaneous expression of MF consists of 4 stages: a patch stage with eczematous features, a plaque stage, a tumor stage, and an erythrodermic stage. Progression to a later stage usually occurs within a few years after the onset of the disease. On the early stage, the color of patches may range from red, purple, brown,

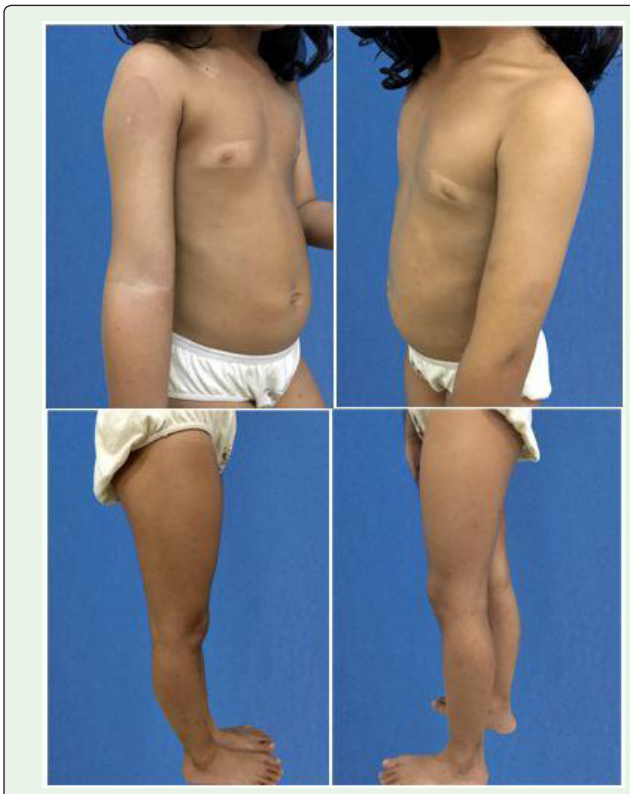

Figure 3: After NBUVB and topical steroid for a year.

to hypopigmented [7,8]. The mechanism of hypopigmentation in MF remains unclear. One of the most acceptable hypotheses is the presence of atypical lymphocytes that infiltrate the epidermis which causes melanocyte degeneration and abnormal melanogenesis as a result of a cell damage. Another theory suggests the possibility of pigment loss due to the defect of melanosome transfer from melanocytes to keratinocytes [5].

The histopathological feature is one of the most important findings to support the diagnosis of MF, but it is usually more pronounced in the plaque or tumor stage. The histopathologic features of early-stage MF are very similar to other skin inflammatory diseases, which show infiltration of inflammatory cells in the superficial layerand around the perivascular. Pautrier's microabscess, epidermotropism of lymphocytes accompanied by a mild or absenceof spongiosis, linearly arranged lymphocytes in the basal layer, as well as larger lymphocytes with hyperconvulated nuclei are the specific findings of MF [1].

Our case showed histopathologic features that imitating leprosy, where we found a collection of mixed inflammatory cells consisting of lymphocytes, histiocytes and plasma cells in the dermis, around the blood vessels and around the nerves. But submitted stains of Ziehl Nielsen and Fite Faraco were negative. We also found epidermotropism of atypical lymphocytes in the absence of spongiosis. These atypical lymphocytic cells are also found around adnexa. The presence of these atypical cells increased our suspicion towards mycosis fungoides, so we proceed to immunochemistry examinations. Immunohistochemical data showed CD20-negative on most cells, CD3-positive on most cells, and positive of CD5, CD4, CD8, CD56, CD30, perforin, and granzyme on some cells.

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CD30-positive is uncommon in MF, but CD30 expression can occur in MF that transforms into anaplastic lymphoma [9], which is found in approximately 40% of cases [10]. CD56 can also be found positive in NK/T cell lymphoma [11]. Therefore, we conducted EBER in-situ hybridization to see the positive Epstein-Barr virus involvement in atypical cytotoxic lymphocyte infiltrates. A positive result supports the diagnosis of NK/T cell lymphoma, which is associated with a poorer prognosis and a poor therapeutic response [11]. Based on the negative findings of EBER, immunophenotyping of leukemia, Sezary cell found, and involvement of lymph nodes or internal organs, the patient was diagnosed as a hypopigmented mycosis fungoides without any systemic involvement.

The treatment approach for childhoodMF is similar to the adult [2,7,8]. The modalities of therapy for hypopigmented MF are topical corticosteroids that can be combined with NBUVB or psoralen and UVA phototherapy [7,8]. Other treatment options believed to be effective are topical nitrogen mustard, topical carmustine, and total skin electro beam therapy [12,13]. However, Boulos et al. would not recommend topical nitrogen mustard as the first-line treatment in childhood MF because of its potential carcinogenic effect [8].

A potent topical corticosteroid can achieve a good response in 94% of patients [8]. In a retrospective study of childhood MF from Turkey, most of the patients treated with topical steroid achieved initial clearance, nevertheless, 69.2% of the patients had recurrences [2]. Until now there is no standard protocol for MF in children, but phototherapy is an effective choice for the treatment of hypopigmented MF types [8]. NBUVB is generally considered to be more effective because it promotes a suppressive effect on systemic immune responses, and is considered less carcinogenic with fewer adverse effects compared to PUVA [2,7,8]. A retrospective review conducted in Singapore shows that all patients treated with NBUVB had at least partial clearance and 49.7% patients achieved complete clearance after a mean duration of 8.9 ± 5.3 months, but 46% of those patients relapsed after a mean duration of 14.9 ± 14.8 months. None of the PUVA-treated patients achieved complete clearance while on treatment [14]. Another retrospective review performed in New York shows 45.7% patients improved with phototherapy but the areas of hypopigmentation did not completely repigment. However 20% recurred after stopping the therapy, and only 7% had complete remission [15].

Our patientwas cleared after one year combination therapy of 0.1% mometasone furoate with NBUVB.We made sure she was cleared by performing a follow-up biopsy and immunochemistry examination. We then tapered down the frequency and dose of phototherapy before we completely stop the treatment. Childhood MF is characterized by frequent relapses which may result in high cumulative doses of UV light, therefore, periodic evaluation is highly important.

Childhood MF has a better prognosis than adults, with a survival rate of 95% in 5 years and 93% in10 years [6]. Delay in diagnosis can lead to progression to a later stage, which negatively affects prognosis. Combination of NBUVB andtopical steroid is a safe and effective treatment for childhood hypopigmented MF.

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