Juvenile-onset non-poikilodermatous CD8+/CD56+ mycosis fungoides

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Background

Childhood cases of cytotoxic T-cell lymphoma have rarely been reported and may be of concern of poor prognosis. We report a very rare case of juvenile non-poikilodermatous C8+/CD56+ mycosis fungoides (MF).

Case presentation

A 22-year-old female presented with a 6-year history of multiple well-demarcated large roundish-oval scaly and reddish-brownish patches and plaques on the trunk and extremities (**Fig. 1**), which developed within about two weeks and remained more or less unchanged since their appearance. Histopathology revealed focal parakeratosis and prominent epidermotropism of atypical lymphocytes (**Fig. 2a**). In the upper dermis, there were scanty eosinophils and brownish melanophages as well. On immunohistochemistry stains (**Fig 2b-d**), the infiltrating lymphocytes were mainly positive for CD8, CD56, and TIA-

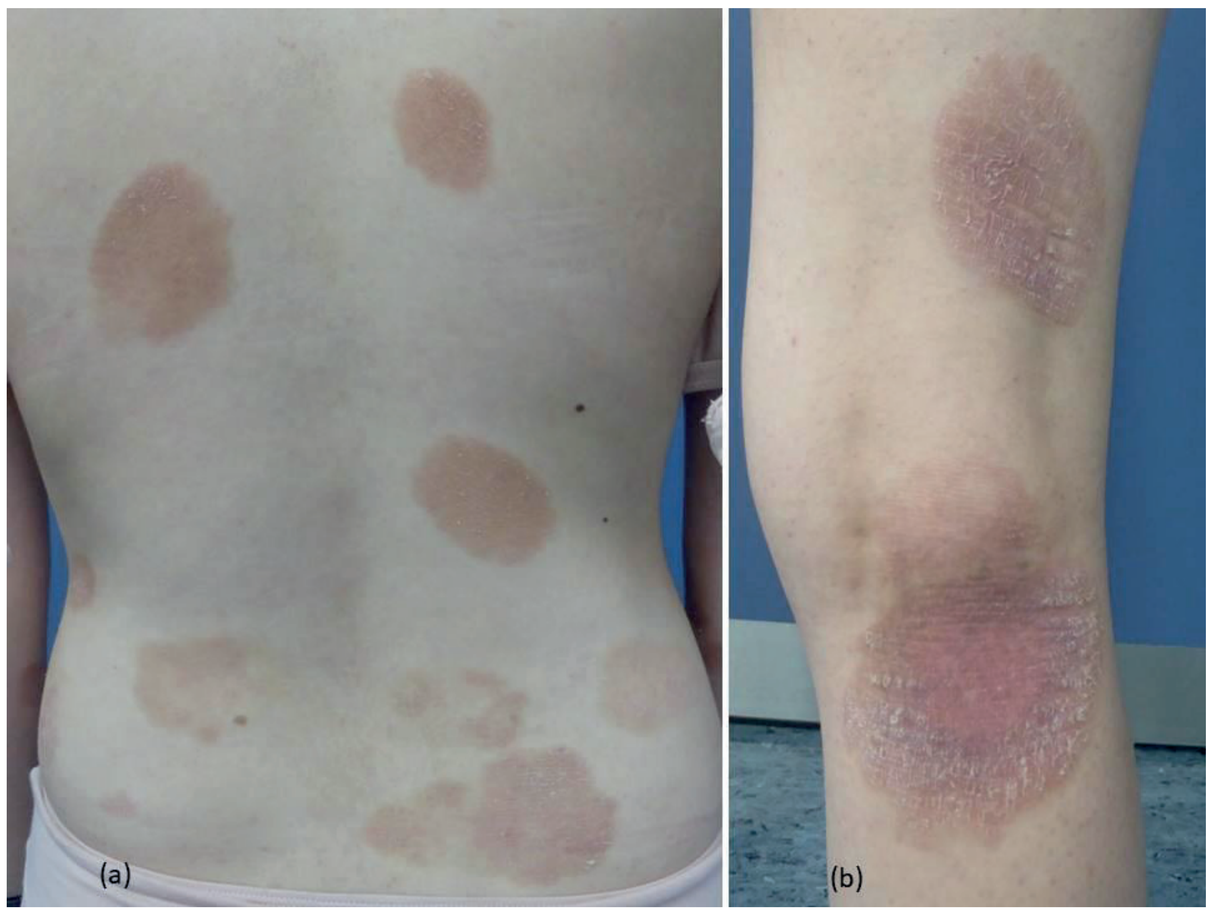

Fig. 1. A young female with well-demarcated scaly reddish-brownish patches/plaques on back (**a**) and dorsal leg (b).

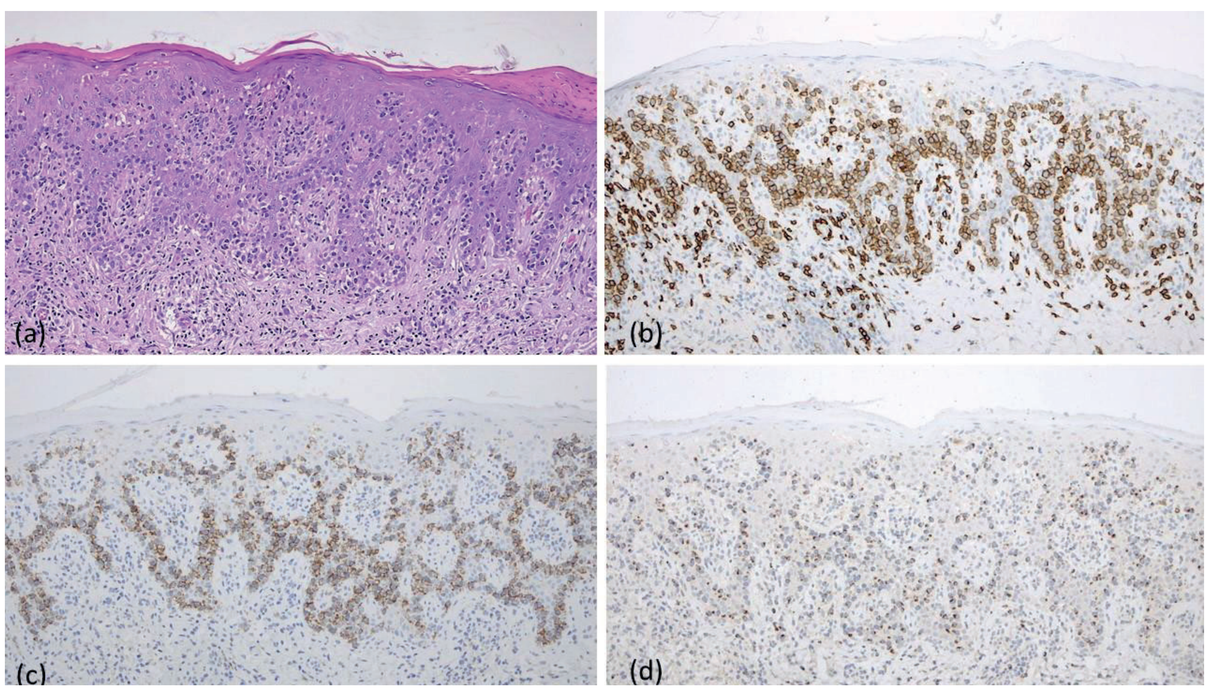

Fig. 2. Haematoxylin-eosin stain of a skin biopsy obtained from the leg (**Fig. 1b**) revealed focal parakeratosis and prominent epidermotropism of atypical lymphocytes. Immunohistochemically (**a**), the infiltrating lymphocytes were mainly positive for CD8 (**b**), CD56 (**c**), and TIA-1 (**d**); magnification: x 100.

and showed loss of CD7 and CD5 expression. CD30 expression was sporadically observed, and if, only in non-atypical T-lymphocytes. T-cell receptor (TCR) gene rearrangement analysis (multiplex-PCR, BIOMED-2) of lesional skin demonstrated the rearrangement of the gamma chain (tube A: 162 nt). Flow cytometry of the peripheral blood revealed slightly decreased CD4/CD8 ratio with an increase of CD8+ cells and decrease of CD4+ cells. A small (max. 0.11%) subpopulation of CD3+ CD8+CD5- cells was found. However, TCR gene rearrangement was not detectable in peripheral blood. Complete work-up including lymph node and abdomen ultrasound and blood smear did not reveal evidence for lymphoma spread. The patient was started on clobetasol propionate 0.05%o ointment and narrowband ultraviolet B phototherapy. The skin lesions markedly improved after 12 weeks of treatment.

Discussion

Uncommon variants of MF have been reported in children and adolescents, particularly including the hypopigmented and poikilodermatous variants. In the present case, a classic clinical MF plaque-type was observed. Immunophenotypically however, the neoplastic T lymphocytes showed strong expression of CD8, CD56, and TIA-1 – a cytotoxic expression profile that is significantly different from classic MF which is characterised by a CD4+immunophenotype. Ben-Amitai et al. reported that the CD8+ MF is overrepresented in the paediatric age group having a very good prognosis. However, the CD8+/CD56+ immunophenotype is extremely rare in children and adolescents with MF. The present case is very similar to the case of a juvenile-onset non-poikilodermatous CD8+/CD56+ MF previously reported by Kempf et al. also presenting with non-poikilodermatous lesions. Most of the previously described patients with this phenotypic variant were adults at the time of diagnosis and often manifested with widespread poikilodermatous skin lesions.

The most important differential diagnosis of juvenile-onset CD8+/CD56+ MF is primary cutaneous aggressive epidermotropic CD8+ cytotoxic T cell lymphoma and subcutaneous panniculitis-like T cell lymphoma which have a much more rapid and aggressive clinical course including early ulcerating and necrotising tumours and subcutaneous lesions, respectively. Since juvenile-onset CD8+/CD56+ MF, primary cutaneous aggressive epidermotropic CD8+ cytotoxic T cell lymphoma, and subcutaneous panniculitis-like T cell lymphoma share common immunohistological features the distinction these conditions must be made predominantly based on clinical features, disease history, and complete work-up results. In the present case, the age at disease onset, clinical work-up, and appearance of skin lesions did not suggest a diagnosis of the above-mentioned aggressive T cell lymphoma subtypes. Another clinical differential diagnosis includes large plaque parapsoriasis which has been described very rarely in childhood. However, histopathology, immunophenotyping, and genotyping aid to differentiate parapsoriasis from juvenile-onset CD8+ MF. Notably, juvenile-onset CD8+ as well as juvenile-onset CD8+/CD56+ MF can be controlled in most patients by skin-directed therapies, including phototherapy and topical corticosteroids.

Conclusions

Juvenile-onset non-poikilodermatous CD8+CD56+ MF represents a very rare MF subtype and is associated with an indolent course. In order to avoid too aggressive diagnostics and treatments, clinicians should be aware of this rare and indolent MF variant in childhood and adolescence.

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