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Pediatric Subcutaneous Panniculitis-Like T-Cell Lymphoma with Features of Hemophagocytic Syndrome

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To the Editor: We read with interest the recently published article by Huppmann et al. [1] describing a cohort of 16 pediatric patients with subcutaneous panniculitis-like T-cell lymphoma (SPLTCL). In their series, they reported only one case with possible hemophagocytic syndrome (HPS). We have encountered another pediatric case of SPLTCL with features of HPS. The patient was a 13-year-old female with a 1-month history of fevers, chills, and progressive 1–3 cm painful subcutaneous plaques and nodules on her face, chest, back, and extremities. Full-body PET scan revealed innumerable lesions involving the trunk and limbs (Fig. 1A). Laboratory evaluation showed leukopenia, anemia, negative antinuclear antibodies and rheumatoid factor, and elevated erythrocyte sedimentation rate, liver transaminases, lactate dehydrogenase, ferritin, and soluble interleukin-2 receptor level.

A skin punch biopsy from the left arm revealed dense infiltrates predominantly confined to the lobules of subcutaneous tissue (Fig. 2A), which displayed a lobular panniculitis-like process consisting of predominantly atypical lymphoid cells rimming around individual fat cells (Fig. 2B). The atypical lymphoid cells were of intermediate size with irregular nuclear contours, hyperchromasia, occasional lobated nuclei, and inconspicuous nucleoli. Increased histiocytes with prominent hemophagocytosis were seen engulfing nucleated cells with associated apoptosis and karyorrhectic debris (Fig. 2C). Scattered plasma cells were also present throughout the infiltrate (Fig. 2D). The atypical lymphoid cells were positive for CD2, CD3, CD7, CD8, and β F1, and were negative for CD4, CD5, CD15, CD30, CD56, TCR γ , and EBV EBER *in situ* hybridization stain (Fig. 2E, F). Monoclonal T-cell receptor beta chain and gamma chain gene rearrangements were detected by polymerase chain reaction. A bone marrow biopsy revealed increased histiocytes with focal hemophagocytosis.

Our case showed characteristics of SPLTCL which is rare in children and is usually described as having an indolent course. However, an association with HPS has been shown to have a more aggressive course and poorer outcome [2]. In the largest series of SPLTCL published so far, which included both pediatric (n = 12) and adult (n = 51) cases, 17% of total patients had HPS [3]; we could not extrapolate, however, if any pediatric cases had HPS. In children, associated HPS appears to be extremely rare, as there was only one possible case with HPS in the series of 16 children by Huppmann et al. [1], and only two other children with HPS have been reported in the Asian population [2]. Of note, the presence of plasma cells, seen in the majority of pediatric SPLTCLs as recently reported [1], was also present in our case, making the differential diagnosis with lupus panniculitis challenging.

Our patient was treated with prednisone followed by hydroxychloroquine and then bexarotene plus prednisone. The response was mixed; some nodules and plaques resolved, while others persisted and new nodules developed with recurrent episodes of fever, chills, night sweats, and persistent leukopenia. A systemic chemotherapy regimen of SMILE (steroids, methotrexate, ifosfamide, L-asparaginase, and etoposide) was then initiated. Follow up full-body PET scans at 6 months and 10 months after initial PET scan revealed resolution of the lesions (Fig. 1B). The patient was alive with no evidence of disease at 24 months after diagnosis.

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Fig. 1. Full body PET image, pre-therapy. There are innumerable FDG-avid nodules and subcutaneous plaques involving the trunk and limbs from below the base of the neck. There are also enlarged FDG-avid bilateral axillary and external iliac lymph nodes (A). Full-body PET image 6 months after the initial PET image, after third cycle of SMILE chemotherapy, showed significant decrease of FDG-avid nodules and subcutaneous plaques (B).

Fig. 2. Histologic findings of the skin punch biopsy from the left arm. Dense, lobular subcutaneous infiltrates are seen with sparing of the epidermis and dermis (A). The infiltrates show a lobular panniculitis-like process with atypical lymphoid cells rimming around fat cells (B). Histiocytes are increased and demonstrate hemophagocytosis, engulfing nucleated cells with associated apoptosis and karyorrhectic debris (C). Scattered increased plasma cells are also

identified (**D**). Immunohistochemical stains demonstrate the atypical lymphoid cells are positive for CD8 (**E**) and β F1 (**F**).