Dermatology Practical & Conceptual

Supplementary Material

Text S1. Course and Outcome.

All patients were referred to the pediatric hematology-oncology department for further screening, including total body imaging and hematologic evaluation.

As no further organ involvement was found, Cases 1, 2, and 3 were diagnosed with a single-system cutaneous LCH. The disease was managed successfully with topical corticosteroids and a watch-and-wait approach; finally, spontaneous remission was observed.

In Case 4, the full-body MRI scan revealed mandibular and ilium involvement, which led to a diagnosis of multisystem, low-risk LCH. The patient received chemotherapy with vinblastine/prednisolone per the LCH-III protocol. Due to a relapsing course, the therapy was switched to MTX/6-mercaptopurine, which has resulted in a current disease remission for nine months thus far.

Case 5 was diagnosed with multisystem, high-risk LCH with cutaneous, intestinal, hepatic, and osseous involvement. The initial therapy according to LCH-III protocol was unsuccessful, with a marked clinical exacerbation under therapy. Because of a confirmed BRAF V600E mutation, the therapy was switched to dabrafenib, which has shown an excellent response, with rapid remission of the lesions in all involved organs.

Case 6 was also diagnosed with a multisystem, high-risk organ LCH with cutaneous, lymphatic, pulmonary, splenic, intestinal, and osseous involvement. The chemotherapy according to the LCH III protocol had initially led to partial disease remission, followed by a relapsing-remitting course. Eventually, a fulminant, therapy-resistant relapse led to fatal outcome due to respiratory failure.

In Case 7, the oncological screening showed additional lesions in the liver, spleen, and cervical lymph nodes, leading to the diagnosis of multisystem high-risk organ LCH. Chemotherapy was initiated according to LCH III protocol. The disease exhibited a remitting-relapsing course, necessitating the re-initiation of chemotherapy. However, long-standing remission was eventually achieved.