Dermatology Practical & Conceptual

Concurrent Langerhans Cell Histiocytosis and Mastocytosis: a Third Known Case

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To the Editor,

We read with interest the recent case report by Boisseau et al. describing an extremely rare association between Langerhans cell histiocytosis (LCH) and mastocytosis, with one reported case in an infant and this first documented case in adults [1,2]. We wish to contribute by presenting, with informed consent, a similar case which further supports their observations.

Our pediatric patient was born full term with a skin rash affecting the whole body except for palms and soles. The rash was described as round, well-demarcated lesions with epidermal defects, varying in size from one to several millimeters, some slightly exudative. Below the right eye there were two raised tumorous lesions measuring 10–15 mm, dry and brown-orange-red in color. Similar skin lesions were also seen on the upper lip, foot, and thorax. Further clinical examination was normal. During the first week of life, the rash faded, and the skin lesions became less pronounced and dried out. Examinations with full-body skeletal X-ray, chest X-ray, abdominal ultrasound, and blood samples were normal. The skin biopsy had positive expression of CD1a, CD68, and S100a, and morphology was consistent with LCH. This congenital skin-limited LCH resolved spontaneously within 2–3 months without any treatment. During the following years, symptoms such as rashes, diarrhea, and ear problems were examined, and repeated biopsies were performed, all of which were normal without evidence of LCH. Magnetic resonance imaging of the brain was performed twice without signs of CNS involvement.

At the age of two months, there were no new LCH lesions. However, a new skin abnormality had developed on the patient's left thigh, occasionally changing in color from barely visible to intensively red, round, 24 x 9 mm lesion

with a small central nodule. The skin biopsy was strongly positive for mast cell tryptase and was diagnosed as mastocytoma. Serum tryptase was normal (4 µg/L) and there have not been any symptoms of systemic mastocytosis.

The patient was followed for ten years by pediatric oncology, without any sign of LCH reactivation nor suspected late sequelae. The cutaneous mastocytoma stayed unchanged, sometimes causing local blistering and itching. This third case on the coexistence of these two hematopoietic neoplasms raises questions concerning the reason for their coexistence, such as the possibility of shared or interacting disease mechanisms [3-5]. Unfortunately, no lesional material from the initial LCH skin biopsy remains available for genetic analysis, which we encourage in any future cases.

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