PAEDIATRIC DERMATOLOGY

DISSEMINATED DE NOVO MYELOID SARCOMA IN A 17-YEAR OLD MALE

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Background: Myeloid sarcoma (MS) is an extramedullary tumour of malignant myeloid blasts. The incidence of MS is 2-9% among all cases of acute myeloid leukemia (AML). The World Health Organization classifies true de novo MS as bone marrow specimens with fewer than 30% blasts on initial evaluation; myeloproliferative diseases frequently develop in these patients. Given the lack of reported cases, treatment protocols are lacking. Disseminated MS remains exceedingly rare, with no pediatric cases reported showing this extensive extramedullary disease. Cutaneous MS is also extremely rare, with limited pediatric case reports in the literature.

Observation: A 17-year old systemically well boy presented with cervical lymphadenopathy and a four-week history of numerous, progressive, asymptomatic, indurated violaceous to red-brown papules and nodules located primarily on his torso, neck and scalp, and with decreased density on his extremities and gingiva. No blasts were seen in the peripheral blood. A moderate-sized anterior mediastinal mass and hilar lymphadenopathy were seen on chest x-ray. Skin and cervical lymph node biopsies were most consistent with AML. After several bone marrow examinations, evidence was ultimately found of limited areas showing leukemic infiltrate, but with 60-70% normal trilineage hematopoiesis. A whole-body PET scan demonstrated diffuse hypermetabolic foci scattered throughout many organ systems. While an initial diagnosis of AML was considered, the lack of significant leukemic bone marrow or blood involvement led to a final diagnosis of de novo MS.

Key Message: Pediatric dermatologists must retain a high index of suspicion for malignant processes, including MS, when faced with a patient with diffuse nodular skin disease. Work-up should include histopathology of skin lesions, including cytogenetic and immunohistochemical analyses, as well as examination of the bone marrow for leukemic infiltration. When absent at presentation, ongoing monitoring for development of myeloproliferative disease is required.