

Juvenile Dermatomyositis and Development of Malignancy: 2 Case Reports and a Literature Review

To the Editor:

In adults, there is an established correlation between dermatomyositis (DM) and malignancy¹, but in children there are very few case reports in the literature. Here we report 2 cases from our institution of development of leukemia after diagnosis of juvenile DM (JDM).

Written consent was provided from the patients (and their parents) included in this report. The Duke University Health System institutional review board (IRB) does not require IRB approval for case reports describing 2 patients.

Case 1. An obese 6-year-old white girl was admitted with weakness and rash over the preceding 6 months with associated weight loss, fever, and joint pain. Family history was negative for malignancy, immunodeficiency, and autoimmunity. On examination, she had symmetric proximal muscle weakness, heliotrope rash, Gottron papules, nailbed telangiectasias, and no evidence of organomegaly. She had an initial Childhood Myositis Assessment Scale (CMAS) of 14/52. Additionally, she had elevated muscle enzymes, muscle edema on magnetic resonance imaging (MRI), as well as electromyography consistent with myositis. She had a positive antinuclear antibody (ANA), negative myositis-specific autoantibodies (MSA) and dsDNA, and normal complements. Myositis-associated autoantibodies (MAA) were not tested.

She was diagnosed with JDM. She received methylprednisolone and intravenous immunoglobulin at diagnosis. She was then transitioned to oral steroids and started on methotrexate (MTX) in addition to hydroxychloroquine. She successfully tapered off oral steroids about a year after diagnosis. Eighteen months after diagnosis of JDM, she developed new onset neutropenia and thrombocytopenia. She underwent bone marrow biopsy and was diagnosed with acute promyelocytic leukemia. She was treated with chemotherapy and remains in remission 8 months after completion of therapy.

Case 2. An obese 16-year-old Hispanic female was admitted with progressive weakness, rash, anorexia leading to a 50-pound weight loss, fatigue, arthralgias, and alopecia over 6 months. Family history revealed prostate cancer in a grandfather but was otherwise negative for malignancy, immunodeficiencies, and autoimmunity. On examination she had a diffuse, erythematous hyperpigmented rash; Gottron papules; and a shawl sign. She had marked alopecia, polyarticular arthritis, and symmetric proximal weakness with a CMAS of 14/52. Her MRI was consistent with myositis. Her deltoid muscle biopsy showed widespread fiber degeneration and regeneration, perifascicular atrophy, and MHC class I upregulation. Computed tomography of abdomen/pelvis was negative for hepatosplenomegaly. Her ANA was positive, dsDNA was negative, complements were normal, MSA testing showed a weak positive melanoma differentiation-associated protein 5, and MAA were negative. At diagnosis she had interstitial lung disease (ILD) that did not require supportive measures. She was treated with methylprednisolone, prednisone, MTX, and rituximab (RTX).

She had a good response to therapy, tapered off steroids, and did not require further RTX treatment. Her ILD went into remission without complication. MTX was changed to mycophenolate mofetil (MMF) because of persistent transaminitis less than twice the upper limit of normal and mild facial erythema. Both resolved within 3 months of MMF initiation.

Thirty-one months after diagnosis, she presented with fatigue, petechiae, easy bruising, and cytopenias; her blood smear revealed blasts. Bone marrow biopsy led to a diagnosis of B cell acute lymphoblastic leukemia. She is currently 3 months into her treatment and doing well.

There are few case reports of children with JDM either having DM as a paraneoplastic manifestation or developing a malignancy after JDM diagnosis. Currently there are no recommendations for routine evaluation of malignancy for children diagnosed with JDM.

We searched for related articles through PubMed (MEDLINE) without restriction on language. Morris and Dare describe 12 patients with

JDM/juvenile polymyositis (JPM) who developed malignancy; 9 of the 12 patients had unusual physical findings at the time of diagnosis such as atypical rash or splenomegaly². All 3 without atypical findings developed malignancy more than 1 year after their diagnosis (13, 36, and 44 months), while the other 9 with atypical findings were diagnosed with cancer within 1 year of their diagnosis of JDM/JPM.

Stübgen's case series described 6 additional patients not included in Morris and Dare's review³. Five of the 6 patients were diagnosed with lymphoma within 13 months after diagnosis of JDM and one of those patients had unusual findings of hepatomegaly at presentation³. All of these cases must be interpreted in the setting of receiving immunomodulatory medications that may influence risk of malignancy⁴.

Though reports of JDM associated with malignancy are rare in the literature^{5,6,7,8,9}, the published case reports do seem to suggest that atypical physical examination findings at diagnosis were more frequently observed in patients who developed malignancy within 13 months of diagnosis of JDM/JPM^{2,3}. Neither of our patients had atypical physical findings at diagnosis and both were diagnosed with leukemia at least 18 months after diagnosis of JDM, which is consistent with existing reports of patients with JDM who develop cancer more than 13 months after JDM diagnosis. The cases we present and existing cases in the literature are a reminder for physicians caring for children with JDM/JPM that cancer is rare but can still occur even years after the diagnosis. However, the presence of atypical features at diagnosis, such as organomegaly, may necessitate closer monitoring for development of malignancy.

LAURA CANNON, MD; JEFFREY DVERGSTEN, MD; CORY STINGL, MD, Division of Pediatric Rheumatology, Department of Pediatrics, Duke Children's Hospital, Durham, North Carolina, USA. Address correspondence to Dr. L. Cannon, Division of Pediatric Rheumatology, Department of Pediatrics, Duke Children's Hospital, 2301 Erwin Road, Durham, North Carolina 27710, USA. E-mail: laura.cannon@duke.edu

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