# Paraneoplastic dermatomyositis in a patient with an oligodendroglioma

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## REPORT OF A CASE

A 26-year-old white man with no significant medical history was admitted for nausea, headache, and rash. Physical examination found multiple, coalescing, erythematous-to-violaceous macules and patches on the eyelids and periorbital regions, cheeks, upper chest (Fig 1, *A*), shoulders (Fig 1, *B*), upper back, and posterior neck. Mild periorbital edema was also found. Review of systems was positive for myalgias. Laboratory evaluation found elevated serum creatine kinase levels (673 U/L); tests for rheumatoid factor, antinuclear antibodies, and anti-SSA and anti-SSB antibodies were negative. Immunoglobulin subclasses were within normal limits, and a myomarker panel, including anti—Jo-1, antitranscription intermediary factor-1-$\gamma$, and antinuclear matrix protein-2 antibodies, was negative. Histopathologic examination of a shoulder lesion found focal vacuolar interface changes at the dermoepidermal junction and a sparse, superficial perivascular dermal lymphocytic infiltrate (Fig 1, *C*). Colloidal iron staining showed dermal mucinosis. Based on these findings, the diagnosis of dermatomyositis was made.

Magnetic resonance imaging of the head found a cerebral cystic mass in the right insula (Fig 2); stereotactic biopsy was consistent with an oligodendroglioma. He received a single 60-mg oral dose of prednisone and topical therapy with triamcinolone 0.1% ointment and hydrocortisone 2.5% ointment. Within 10 days, the rash completely resolved. He underwent a craniotomy for tumor resection several weeks after initial presentation. At follow-up 4 months later, he showed no recurrence of myalgias or rash.

## DISCUSSION

Dermatomyositis is a multifactorial inflammatory myopathy involving the integumentary and muscular systems. It may occur as either an idiopathic or paraneoplastic phenomenon; in the largest cohort study to date, Chen et al[1] found a 9% incidence of malignancy among patients with dermatomyositis. Risk of malignancy is highest in patients older than 60 years and remains elevated for at least 5 years; it is controversial how long patients should undergo increased surveillance after diagnosis.[2] Although numerous malignancies have been associated with dermatomyositis, ovarian, lung, and gastrointestinal cancers are the most common in the Western world.[3]

To our knowledge, only 2 previous cases of dermatomyositis associated with an intracranial neoplasm have been reported. One case occurred in a 39-year-old man following dendritic cell immunotherapy for an oligoastrocytoma.[4] The second case was that of a 7-year-old girl with a choroid plexus papilloma.[5] Our case illustrates dermatomyositis in a patient with an oligodendroglioma.

The mainstay of treatment for all dermatomyositis patients is systemic immunosuppression with corticosteroids. As in our patient, topical corticosteroids and calcineurin inhibitors may also be helpful. In paraneoplastic cases, treatment of the underlying malignancy may result in the elimination of symptoms, which may, however, recur if the cancer returns.[3]

Although some patients have positive serology for a variety of biomarkers of paraneoplastic phenomena, other patients, like ours, may be

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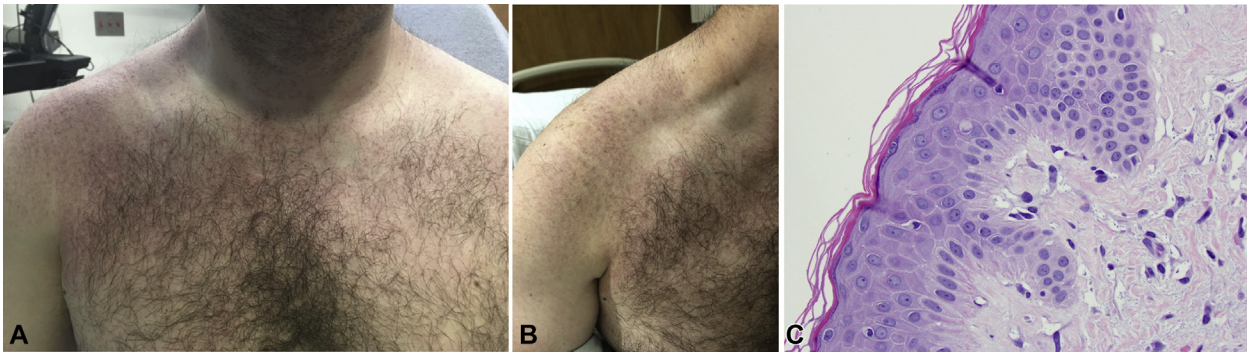


**Fig 1.** **A**, Violaceous patches on the upper chest. **B**, Erythematous-to-violaceous macules and patches on the upper shoulder. **C**, Biopsy shows focal vacuolar interface changes at the dermoepidermal junction and sparse, superficial lymphocytic infiltrate.

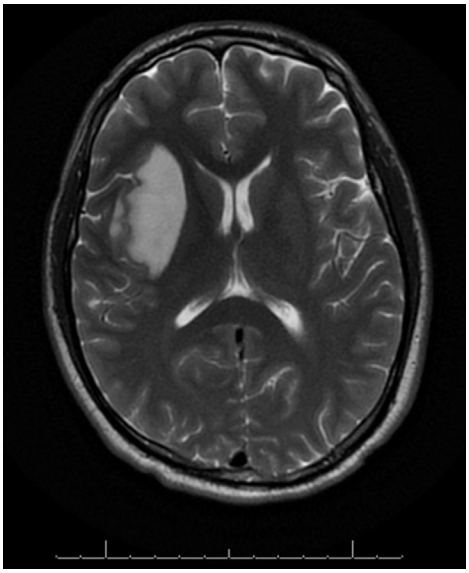

**Fig 2.** Magnetic resonance imaging shows a mass in the right insula.

antibody negative. Newer assays for dermatomyositis-associated autoantibodies have broadened the range of diagnostic tests available to help identify patients likely to harbor occult malignancies. For example, antitranscription intermediary factor-1-γ antibody and antinuclear matrix protein-2 antibody have been associated with an increased risk of malignancy in dermatomyositis patients older than 45 years.[6] However, in a study of 213 patients with dermatomyositis, only 55% tested positive for either of these antibodies, and the utility of this assay has yet to be reported in children or young-adult patients.[6]

Given the morbidity and mortality associated with many cancers, it is imperative for clinicians to recognize dermatomyositis as a possible harbinger of malignancy. This report adds oligodendroglioma to the list of potential neoplasms to be considered when assessing a patient with dermatomyositis.

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