E-Poster

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An Extremely Rare Manifestation of Severe Dermatomyositis: Edematous and Erythematous-Squamous Form - A Case Report

Topic: Diagnosis

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BACKGROUND

Edematous and erythematosquamous dermatomyositis (DM) is an extremely rare clinical subtype of dermatomyositis, distinguished by localized or widespread subcutaneous edema and erythematosquamous skin lesions. It is typically a severe form, often requiring prompt therapeutic intervention. Our case highlights the diagnostic and therapeutic challenges associated with this uncommon variant.

CASE DESCRIPTION

A 37-year-old woman was hospitalized for DM presenting with atypical skin and muscle involvement. Her symptoms included diffuse edematous and erythematosquamous lesions predominantly affecting the upper limbs, along with proximal muscle weakness and myalgia. She was initially treated with methylprednisolone boluses followed by prednisone. However, her condition worsened, with increased edema, progression of cutaneous and muscular involvement, and the onset of swallowing difficulties. Intensifying treatment with azathioprine led to the resolution of cutaneous symptoms, including edema, along with muscle strength recovery and improvement of dysphagia.

DISCUSSION

In our patient, the presence of diffuse edematous and erythematosquamous skin lesions, along with proximal muscle weakness and progressive dysphagia, aligns with the clinical features reported in the literature. Some studies have emphasized that this DM variant is often refractory to corticosteroid monotherapy, requiring the early addition of an immunosuppressant. The initial worsening of symptoms under corticosteroid therapy alone required the addition of azathioprine, which led to an improvement in both cutaneous and muscular symptoms, highlighting the importance of an aggressive therapeutic approach in this severe form. Other reported cases have shown variable efficacy of immunosuppressants such as azathioprine, methotrexate, mycophenolate mofetil, as well as biologic therapies like rituximab in corticosteroid-resistant cases. The favorable outcome observed with azathioprine in our case suggests that this drug could be an effective therapeutic option for this rare DM variant. However, long-term follow-up remains essential to monitor potential relapses and adjust treatment accordingly. Thus, this case highlights the need for early diagnosis and appropriate management to prevent systemic complications, particularly dysphagic involvement, which can be life-threatening.

CONCLUSION

Subcutaneous edema, whether focal or widespread, should not be confused with periorbital edematous erythema, a hallmark feature of dermatomyositis. This type of edema may indicate disease severity, frequently linked to pronounced muscle impairment and swallowing disorders. In such instances, prompt and intensive management combining corticosteroid and immunosuppressive therapy is crucial.

Statement of Disclosures

None

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