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Dermatomyositis Associated with Hemophagocytic Syndrome: A Case Report/Review of the Literature

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Abstract: Hemophagocytic syndrome (HPS) is a disorder that might be transferred genetically due to an autosomal recessive genetic defect of the long arm of chromosome 9 and chromosome 10 (9q21.3-22 and 10q21-22), termed as primary hemophagocytic syndrome (PHPS) or familial hemophagocytic hemophagocytosis (FHPS) and it may be associated with a variety of infections, malignant neoplasms, drugs, autoimmune diseases and various immuno deficiencies, termed as secondary hemophagocytic syndrome (SHPS). For most patients with HPS, the outcome is rapid and fatal unless the diagnosis is made early and followed by prompt therapeutic intervention. Fever, hepatosplenomegaly, cytopenia, hypertriglyceridemia and/or hypofibrinogenemia, low erythrocyte sedimentation rate, hyperferritinemia, hyponatremia and hemophagocytosis shown in bone marrow, lymph nodes or spleen biopsy specimens constitute the clinical presentations of the syndrome. In this paper, a patient diagnosed with dermatomyositis associated with HPS is reported with the clinical findings of fever, lymph node enlargement, weakness and atrophy of proximal muscles, periorbital edema, skin thickness and symmetric violet erythema of the forearms. To our knowledge only two cases of dermatomyositis associated with HPS have been published in the medical literature. In addition, periorbital edema together with dermatomyositis is a very rare condition in the literature.

Key Words: Dermatomyositis (DM), hemophagocytic syndrome (HPS), periorbital edema

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