Abstract

Dermatomyositis (DM) is an immune-mediated myopathy, characterized by proximal muscle weakness and evidence of muscle inflammation along with pathognomonic skin manifestations, including periorbital heliotrope eruption and Gottron papules. DM is known to cause cutaneous and systemic vasculopathy, which is considered central to the pathogenesis. Dynamic changes in vascular lesions play an important role in the occurrence and development of DM. Many studies have documented that vasculopathy is a predominant feature of DM. Here, we report a 48-year-old female who was diagnosed to have DM following her presentation with livedoid vasculopathy with other multiple vasculopathic manifestations.

Case presentation

A 48-year-old lady presented with acute onset, severe, burning pain over right hand and left foot, cold extremities, healed Gottron's papules, livedo racemosa over bilateral lower limbs, over proximal muscle groups, elvated blood pressure and blood pressure difference between bilateral arms. Erythrocytese dimentation rate (ESR) was mildly elevated with normal C-reactive protein(CRP), and creatine kinase (CK) was significantly high, nerve conduction test and electromyography findings were suggestive of mononeuritis multiplex and associated myositis or myopathy. Skin biopsy of livedoid rash suggested vasculopathy with thrombosis. Diagnosis of dermatomyositis was made. She was started on intravenous (IV) cyclophosphamide pulses followed by oral prednisolone along with anti-hypertensives. She exhibited a favorable response to the above treatment and is currently in remission.

Conclusion

Vascular involvement in DM is always under-evaluated due to its diversity ofclinical manifestations. Delayed treatment is associated with poor prognosis withlethal complications. The long-term outcome is better achieved with earlyaggressive therapy. This case report highlights the importance of early identification of vasculopathic manifestations and early aggressive treatment inachieving early disease remission and better prognosis.