A case of clinically amyopathic dermatomyositis with facial edema as a sole cutaneous manifestation

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In clinically amyopathic dermatomyositis, the hallmark cutaneous manifestations are the key to the diagnosis. We report a case of clinically amyopathic dermatomyositis limited to the orbital and masticator muscles that presented with facial edema as the sole cutaneous manifestation and posed a diagnostic and therapeutic challenge. A 58-year-old woman presented with edema of the face that had developed about one year ago. There was no weakness in extremities, and serum creatine kinase level was within normal range. On MRI, there was diffuse edematous change in the bilateral masticator and extra-ocular muscles, accompanied by subcutaneous fat infiltration in the face. On PET-CT scan there was diffuse F-18-FDG uptake in the periorbital, frontalis, temporalis and masseter muscles, and the buccal and submandibular spaces without any increased uptake in extremities. Head and neck surgery, and radiology department gave opinions that a biopsy of the muscles would be risky. Therapy with medium-dose glucocorticoid in combination with either azathioprine or mycophenolate or methotrexate was not successful in inducing clinical remission, and the facial edema relapsed in days at glucocorticoid doses lower than 15 mg/day. Then, US-guided core-needle biopsy with an 18-gauge needle was performed in the right masseter muscle. Two pieces of tiny muscle tissues measuring up to 0.5 x 0.1 cm were obtained. On pathologic examination, there were patchy CD4+ T cell-dominant lymphoplasmacytic infiltration in t he stroma, necrosis of myofibrils and prominent perifascicular atrophy. Based on those findings, a diagnosis of clinically amyopathic dermatomyositis was made. Therapy with gamma-globulin was initiated, and by fourth weeks, the facial edema had resolved completely. To the best of our knowledge, it is the second reported case of clinically amyopathic dermatomyositis presenting with isolated facial edema. In consistent with previous reports that showed that the subcutaneous edema is a severe manifestation of dermatomyositis, she needed gamma-globulin therapy to control her disease activity, although the disease extent was limited to the head.