Facial and Laryngeal Edema in a Patient with Dermatomyositis

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The patient, a 47-year-old woman, presented with a 1-month history of hoarseness, mild dysphagia, proximal muscular weakness and a heliotropic rash. Localized face and neck edema was evident (Picture A). The level of creatinine phosphokinase was elevated at 4,193 U/L (normal range: 45-163 U/L). The clinical, laboratory and biopsy findings were suggestive of dermatomyositis. Fat-suppressed, T2-weighted MRI showed laryngeal edema and extensive fluid collection in the adjacent subcutaneous layer (Picture B). In addition, severe laryngeal edema was confirmed on endoscopy (Picture C). The absence of other conditions that can cause edema, including angioedema/allergies, hypothyroidism, deep vein thrombosis and malignancy, strongly suggested that the edema was most likely caused by the pathophysiology of dermatomyositis. The simultaneous occurrence of inflammatory myopathy and localized facial and laryngeal edema is extremely rare (1). However, this combination represents a unique feature that distinguishes such variants of inflammatory myopathy from typical cases.

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Reference