Facial and oropharyngeal angioedema with rash: A rare and interesting clinical presentation of systemic lupus erythematosus

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Systemic lupus erythematosus (SLE) is a systemic autoimmune illness characterized by damage to cells of the body due to autoantibodies and immune complexes formation. SLE affects females more than males (9:1) [1]. In up to 15% of cases, they present with cutaneous manifestations in forms of discoid lupus, subcutaneous cutaneous lupus erythematosus, and erythema multiforme. Angioedema is an area of swelling of the lower layer of skin and tissue just under the skin or mucous membranes [2]. The swelling may occur in the face, tongue, larynx abdomen, or arms and legs. Onset is typically over minutes to hours. Based on mediator, it can present over minutes (histamine induced) or days (bradykinin induced) [3].

An 18-year-old female presented to the outpatient department with complaints of diffuse swelling of face and discoloration along with hoarseness of voice for 5 days. Initially, it involved only the area around her lips but has progressed and involved her whole of the face (Fig. 1). She also gives a history of non-specific complaints of low-grade fever which was not associated with chills and rigors for which she took nonsteroidal anti-inflammatory drugs. On examination, the patient has diffuse swelling of the face along with a reddish-brown non-photosensitive macular rash. The patient also had swelling of mucous membranes and soft tissue of mouth with diffuse vocal cord edema on laryngoscopy. She was treated with nebulization with racemic epinephrine and subcutaneous epinephrine 0.5 mg IM overdose (OD) for 5 days which relieved her difficulty in breathing; however, her swelling persisted. On investigation, her complete blood chemistry, liver function test, and renal function test were normal. The patient was evaluated for autoimmune disorders and her antinuclear antibody 4+ and anti-double-stranded DNA was 156 IU The patient was diagnosed with acute cutaneous lupus erythematosus and was started on hydroxychloroquine 300 mg OD and steroids 60 mg/kg.

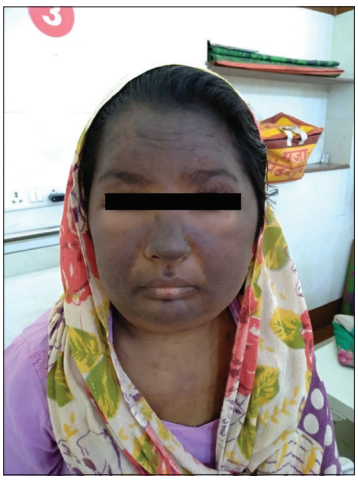

Figure 1: Facial angioedema with rash

Her respiratory complaints completely subsided while the rash faded but yet persisted over the malar region.

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