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Clinical Images

Mucocutaneous blisters & a mediastinal mass: Lifesaving role of surgery

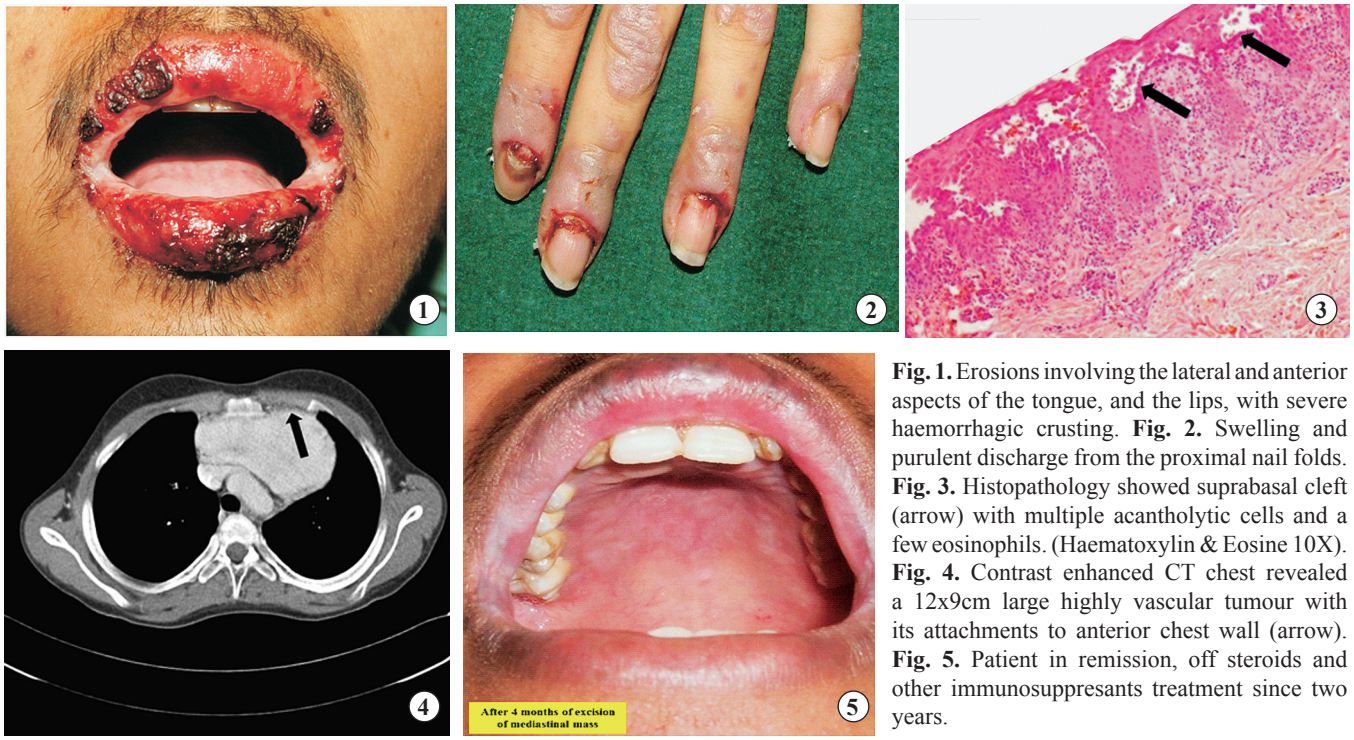

Fig. 1. Erosions involving the lateral and anterior aspects of the tongue, and the lips, with severe haemorrhagic crusting. **Fig. 2.** Swelling and purulent discharge from the proximal nail folds. **Fig. 3.** Histopathology showed suprabasal cleft (arrow) with multiple acantholytic cells and a few eosinophils. (Haematoxylin & Eosine 10X). **Fig. 4.** Contrast enhanced CT chest revealed a 12x9cm large highly vascular tumour with its attachments to anterior chest wall (arrow). **Fig. 5.** Patient in remission, off steroids and other immunosuppresants treatment since two years.

A 16 years old patient presented at Department of Dermatology & Venereology, All India Institute of Medical Science, New Delhi, India, in December 2010 with persistent painful erosions of buccal mucosa and tongue, haemorrhagic crusting of lips and vesiculo-bullous skin lesions (Fig. 1). The proximal nail folds showed swelling, purulent discharge and lichenoid skin lesions (Fig. 2). On Tzanck smear multiple acantholytic cells were seen and histopathology was consistent with pemphigus vulgaris (Fig. 3). Desmoglein (DSG) 1/3 ratio was 18.4/180 µg/ml. A mediastinal mass was incidentally found on chest X-ray, which on contrast enhanced computed tomography appeared as heterogenous contrast enhancing lesion of 12 × 9 cm size in anterior mediastinum (Fig. 4). On angiogram, the lesion was highly vascular with its supply from infra thoraco-cervical trunk and aortic arch. Therapeutic embolization was performed and after six weeks and the patient was taken up for surgical excision of the residual lesion which ultimately turned out to be Castleman's disease. The disease showed remission with minimal immunosuppressive therapy within four months of excision of mass and no recurrence was noted on follow up for two years without treatment (Fig. 5). An underlying Castleman's disease (a rare lymphoproliferative disorder) should be looked for in

cases of pemphigus with atypical features, as it is easily treatable and the treatment can bring about regression of cutaneous symptoms.

Sunil K. Kothiwala & Neena Khanna*
Department of Dermatology & Venereology,
All India Institute of Medical Sciences,
Ansari Nagar, New Delhi 110 029, India
**For correspondence*:
neena_aiims@yahoo.co.in

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