Letters to the Editor

Involvement of upper esophagus resulting in aspiration has been reported. Myocarditis and cardiac dysfunction have occurred in one patient. Our patient had involvement of vocal cord as evidenced by videolaryngoscopy with inter arytenoids thickening. Decreased motility in upper part of esophagus was observed in upper gastro-intestinal endoscopy. The involvement of vocal cord has lead to hoarseness of voice, and it was successfully treated with methyl prednisolone. Even after extensive searches of literature, we have found that vocal cord involvement in scleredema is unreported till date. As of our knowledge, this is the first case of scleredema adultorum of Buschke with involvement of vocal cords.

The treatment protocol for scleredema adultorum has not been effectively described in the literature. Steroids and immunomodulators have been tried for systemic involvement of this disease. Scleredema, often thought to be a benign disease, can rarely present with medical emergencies as was the case with our patient. Patients with scleredema have to be screened for internal organ involvement to prevent mortality.

Kavitha Mohanasundaram, Subramaniyan Kumarasamy, Ramesh Kumar, Chandran P. Rajendran

Departments of Internal Medicine, and *Rheumatology, SRM Medical College and Research Centre, SRM University, Kattangaluthur-603203, India

Address for correspondence: Dr. Kavitha Mohanasundaram, B 404, Staff quarters, SRM Medical College and Research Centre, SRM University, Kattangaluthur-603203, India. E-mail: mmkavitha.96@gmail.com

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returned to us with a similar complaint. Examination showed a well-defined erythematous, firm, shiny, non-tender plaque (7 × 5 cm) on the extensor surface of her left forearm. There was relative clearing and fine scaling at the center of the lesion. The margins of the scales were attached peripherally [Figure 1b]. A lesion with similar morphology was also present on her right shoulder (2 × 3 cm) [Figure 1c].

There was no regional lymphadenopathy. There was gradual central clearing and peripheral expansion with fine trailing scales [Figure 2].

The complete hemogram of the patient was normal, except for the raised ESR (63 mm/hr). Biochemical panel was normal. Skin scrapings for dermatophytes did not reveal any fungal element. Mantoux test (5 TU) showed an induration of 12 mm × 14 mm. ANA (by immunofluorescence; Hep2 cell line) was negative. Chest X-ray, ultrasonography of the abdomen, and gastrointestinal endoscopy was non-contributory. Histopathological examination (from both forearm and shoulder lesions) showed normal epidermis without any spongiosis or parakeratosis. There was papillary dermal edema with slightly dilated blood vessels, and dense perivascular infiltrate, mainly comprising of neutrophils, mononuclear cells and histiocytes. There were extravasations of RBCs, leukocytoclasia, and nuclear dusts formation without any evidence of vasculitis [Figure 3]. The subcutaneous tissue was normal. Direct immunofluorescence was negative.

Based on the clinical and histopathological picture, a diagnosis of recurrent neutrophilic figurate erythema was made. The patient was treated with emollients, topical fluticasone propionate ointment (0.005%) for 2 weeks, and dapsone (100 mg/day). The lesions completely resolved uneventfully within 8 weeks. The dose of dapsone was gradually tapered and stopped at 12th week. There was no recurrence of lesions during a follow-up period of 1 year.

Only a few cases of NFE have been described in adults. Table 1 summarizes reported cases of NFE in adults in the English language literature as revealed by a PubMed database search.\[2,4,5\]
The lesions of NFE are asymptomatic or mildly pruritic and are characterized by arciform plaques, which may have a raised, firm erythematous border and trailing scale. Histologically, the lesions show superficial and deep perivascular and interstitial infiltrate of neutrophils and nuclear debris, as seen in our patient. Other clinical differential diagnoses that were considered in the present case were, erythema annulare centrifugum (EAC), erythema chronicum migrans (ECM), erythema gyratum repens (EGR), annular variant of subacute cutaneous lupus erythematosus (SCLE) and Sweet syndrome. These differentials were ruled out on clinical and histopathological grounds.

Other histological differential diagnoses that present with neutrophilic dermatosis, like, pyoderma gangrenosum, rheumatoid neutrophilic dermatosis, Behcet’s syndrome, urticarial vasculitis, and acute generalized pustulosis, could easily be ruled out by the absence of the distinctive clinical features characteristic of these conditions.

NFE may be present with some underlying disease or in otherwise healthy individuals. One case of NFE was described in an old man as a paraneoplastic manifestation of underlying malignancy. Another case of neutrophilic figurate erythema with the morphology of erythema gyratum repens was reported in a patient with S.L.E. However, our patient did not have any such association.

History of recurrence of lesions on more or less the same site within a span of a few months was an interesting feature in our patient. We seek to emphasize by this report that in spite of its rarity, a diagnosis of NFE should be considered in the differential diagnosis of erythematous scaly annular plaques in adults. Furthermore, while dealing with an annular or figurate inflammatory dermatosis in an adult, the treating physician should exclude any underlying disease including malignancy.

**Table 1: Neutrophilic figurate erythema in adults: clinical summary**

| Reference          | Sites of involvement                         | Age (years) at presentation/gender (Female = F, Male = M) | Underlying disease                                      | Number of episode | Course                                                                 |
|--------------------|----------------------------------------------|----------------------------------------------------------|---------------------------------------------------------|-------------------|------------------------------------------------------------------------|
| Özdemir M et al.   | Back, extremities, and breasts               | 39 / F                                                   | None                                                    | 1                 | Treated with colchicine and hydroxyzine. The lesions resolved uneventfully within a month. No recurrence within 6-months follow-up period. |
| Tre´bol I et al.   | Upper and lower limbs, including the palms and the soles | 79 / F                                                   | Cryptogenic hepatic cirrhosis and Hodgkin’s lymphoma    | 1                 | Complete remission of the lymphoma with radiotherapy. No recurrence of the cutaneous lesions within a 2-year follow-up. |
| Khan Durani B et al. | Trunk, leg                                | 69 / M                                                   | Systemic Lupus Erythematous                             | 1                 | Erythema and infiltrated plaques disappeared after 2 weeks of treatment with local steroids. The patient received cyclophosphamide for lupus nephritis. No relapse was noted during follow-up. |
| Present case       | Forearm, shoulder                           | 47 / F                                                   | None                                                    | 3                 | Dapsone, emollients, and topical steroid caused complete regression of the lesions within 6 weeks. No further recurrence within a follow-up period of 1 year. |

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