Unilateral Mucocutaneous Lymphangioma Circumscriptum of
the Face: Treated Successfully with Sclerotherapy and Laser

Sir,

Lymphangioma circumscriptum (LC) is very rare
hamartomatous, congenital malformed growths of the
lymphatic system in skin and subcutaneous tissue. In
the classification of lymphangiomas, LC has been placed in
the superficial subgroup under the deep (macrocystic) group.
They have classical clinical and histopathological features
such as presence of translucent, discrete or group vesicles
(frog spawn), and by grouped cystic spaces in the papillary
dermis.[1] Herein, we present a rare case of mucocutaneous
LC, involving right oral mucosa and chin skin and treated
successfully with sclerotherapy and laser ablation. This is
the first case report in world literature as per our knowledge.
A 15-year-old boy presented with growth in the right buccal
mucosa extending to chin skin since childhood. The lesion
interfered with mastication, impinged between teeth causing
pain and rarely bled. The chief complaint, however, was cosmetic appearance as it involved a visible portion of oral cavity and face skin. Clinical examination revealed linear, multivesicular swellings and hemorrhagic, crusted swellings (red to blue in color) extending up to angle of mouth [Figure 1a]. These tumors were nontender, nonpulsatile with an irregular warty surface and firm in consistency. The right side of the chin skin showed grouped vesicles with flesh colored to dark red and black colored [Figure 1b]. A clinical diagnosis of lymphangioma was made due to frog spawn appearance, after which biopsy from mucosa was taken. Radiological investigations are such as color Doppler (CD) and magnetic resonance imaging (MRI) scanning. CD studies showed soft-tissue swelling with echogenic character and mild vascularity, but MRI showed hyperintense lesion involving right lip and buccal mucosa; indicating a risk of relapse [Figure 2a and b]. Routine blood investigations were normal.

Histopathological examination revealed grouped dilated cystic spaces in the papillary dermis. The cystic spaces were lined by endothelial cells containing blood and lymphatic fluid at places [Figure 3]. We finally made a diagnosis of unilateral mucocutaneous LC.

As our patient had a history of relapse postexcision from a local general surgeon 5 years back, he was reluctant for any surgery. Thus, we decided to go for sclerotherapy injections for oral lesions and laser ablative surgery for skin growths. We injected Sodium tetradecyl sulfate solution 3% (Sterol) diluted in 30 ml normal saline subcutaneously with an insulin syringe. This was repeated after 2 months. Lesions reduced dramatically after the first session, and there was total clearance after second sclerotherapy session. For the skin on the surface of the chin, two passes of erbium 2900 nm 3 mm during the second visit almost cleared the growths [Figure 4a and b]. We wanted to utilize two different modalities and compare outcome for our case, which was a satisfactory one. There was no relapse in the past 10 months of regular follow-ups.

**Discussion**

LC is a term best reserved for a lymphatic malformation that is localized to an area of skin and subcutaneous tissue. These lymphatic malformations account for 4% of all vascular malformations and about 25% of all vascular tumors and present early in life (0–2 years), mostly in the head and neck area. Mucocutaneous LC (MCLC) is a very rarely encountered in clinical practice. To the best of our knowledge, there has been no reported case of MCLC in world literature, other than the report on zosteriform LC on right upper back skin.[3] LC was first described by Fox and Fox in 1879 under the name lymphangiectodes, the present term was given by Morris in 1889, and its dermatopathology was described by Whimster in 1976.[1,2]

These malformations have to be differentiated from vascular malformations such as pyogenic granuloma, hemangiomas, teratoma, neurofibroma, viral warts, molluscum papules, condyloma growths, herpes infection, and cutaneous metastatic growths (carcinoma telangiectodes).[2,3]

Incisional biopsy for histological examination remains the most definitive diagnostic tool for oral lymphangioma. Radiological studies such as CD, computed tomography (CT) scanning, MRI
studies help to know the location and extension of the growth as well as the surrounding structure. This helps in planning the treatment modality – medical, surgical or combination, which was used in our case. These studies help in assessing the chances of relapses also.\textsuperscript{[4,5]}

Many treatment modalities are available for these rare benign growths such as incision and drainage, cryotherapy, radical surgical excision, radio cautery, laser ablative surgery, sclerotherapy, oral sildenafil, and sirolimus.\textsuperscript{[4,6,7]}

As these growths have a high tendency of recurring after surgical excision, because of their deep component, they are best managed conservatively. Sclerotherapy is the mainstay of treatment of macrocystic lymphatic malformations, particularly a case like ours.

In conclusion, we describe a rare variant of LC, namely MCLC involving the right side of chin and oral mucosa in a 15-year-old male, who showed a remarkably favorable response to sclerotherapy-laser ablation treatment, as manifested by clearance of the growths without scarring or relapse in 9 months of follow-up. However, a long-term follow-up is necessary, and further studies are required to establish the efficacy of sclerotherapy as these cases are known for relapse.

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Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

Mahesh M Unni, Khandelwal Ankit, Sriteja Devalla, Vishakha Chandele
Department of Dermatology and Leprology, MIMSR Medical College, Latur, Maharashtra, India

Address for correspondence: Prof. Mahesh M Unni, Department of Dermatology and Leprology, MIMSR Medical College, Ambajogai Road, Latur - 413 512, Maharashtra, India.
E-mail: pm2Unni@gmail.com

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