

IMAGES

Squamous Cell Carcinoma on the Upper Lip of a Patient with Discoid Lupus Erythematosus

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Discoid lupus erythematosus (DLE) is a chronic autoimmune mucocutaneous disease with an unknown etiology. Typically, central atrophy, small white keratinized plaques with elevated borders, and telangiectasia are seen in patients with DLE. Lesions are found on the oral cavity in 20% of patients with DLE, and upper lip involvement occurs in less than 3% of the oral cavity lesions [1,2]. Squamous cell carcinoma (SCC) developing in a lesion of DLE is very rare, and

is extremely rare on the upper lip [2].

A 49-year-old male patient with a recurrent oral ulcer had been diagnosed with DLE 9 years earlier. Multiple recurrent crusted ulcerative plaques had developed along the upper lip vermilion 7 years earlier. A protruding mass lesion had occurred at the right upper lip 1 year before presentation to our institution, and a wedge resection of the lesion was performed promptly. The lesion was confirmed to be verruca vulgaris. On serial follow-up, the fast growing mass that was confirmed as SCC in the left upper lip had a concurrently expanding crusted lesion, which was confirmed to be keratoacanthoma in the right upper lip. The affected area of the carcinoma was about 4.0 × 2.2 cm, horizontally occupying about 70% of the upper lip, and vertically occupying 80% (Fig. 1). Several enlarged lymph nodes of both submental areas were found using preoperative magnetic resonance imaging. A full thickness defect of 5.5 × 2.5 cm had developed after a wide excision, and bilateral functional cervical lymph node dissection was performed by the head and neck surgery team (Fig. 2).

A bilateral nasolabial orbicularis oris myocutaneous flap was designed and elevated adjacent to both sides of the upper lip defect, along the course of the angular vessel and the remaining orbicularis oris muscle. The near-total upper lip defect was covered successfully by rotation and advancement of the island flap. The remaining mucosal defect was covered with a full-thickness skin graft from triangularly shaped skin, which was located on the lateral side of the alar base (Fig. 3).

There were no significant postoperative compli-



Fig. 1.
Photograph of a 49-year-old male discoid lupus erythematosus patient. A 4.0 × 2.2 cm squamous cell carcinoma on the left upper lip (red arrow) and keratoacanthoma on the right upper lip (blue arrow).



Fig. 2.
Photograph of the lip defect after wide excision of the squamous cell carcinoma. A near-total upper lip defect developed, and the flap was designed on the left side first.

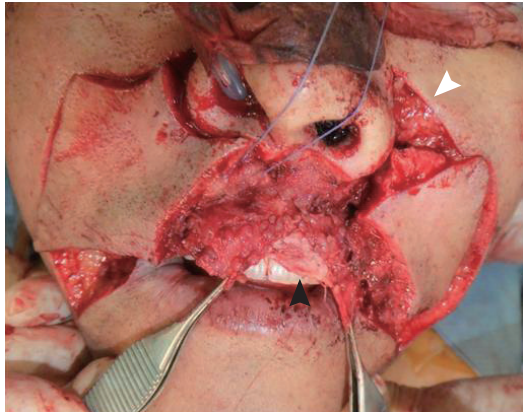


Fig. 3.

Photograph after elevation of the bilateral flaps. The bilateral flap was elevated with a suture placed on the orbicularis oris muscle. A full-thickness skin graft (black arrow-head) from triangularly shaped skin, which was located on the lateral side of the alar base (white arrow head), was used for coverage of the oral mucosal defect.



Fig. 4.

Photograph at postoperative 2 weeks. Oral competence was excellent, and there was no symptom of microstomia.

cations. Oral competence was excellent and sensation was preserved. There was no microstomia, and the patient was satisfied with the results (Fig. 4).

SCC is a rare complication of long-standing DLE. The overall incidence of SCC in DLE has been reported to be 3.3% [1]. The lower lip has been the most commonly affected area in DLE and DLE-related malignant transformation, while the upper lip has been affected in 2.3% of DLE-related SCC [1,2]. It has been reported that ultraviolet irradiation, chronic infection, scars, and long-term immunosuppressive status are possible predisposing factors for the development of SCC in DLE [1-4].

The diagnosis of SCC can be clinically suggestive, but it is essential for SCC to be histologically confirmed. Asanafi and Werth [4] suggested that a diagnosis should only be made after repeated biopsies and careful reevaluation of the course of a specific lesion in relationship to the lupus skin lesions in a patient because differentiating between hypertrophic DLE and SCC can be difficult both clinically and histologically. Our patient was also histologically diagnosed with verruca vulgaris before diagnosis of SCC.

DLE-related SCCs have been observed to be more aggressive than conventional SCCs [1]. The recurrence, metastasis, and mortality rates were 10% to 20% higher than that of non-DLE-related SCCs [3]. Therefore, aggressive therapy is warranted, and a near-total upper lip defect is an inevitable consequence.

The reconstructive method for the upper lip should be chosen in consideration of the orbicularis sphincter

function and aesthetic outcomes. Considering these factors, the nasolabial orbicularis oris myocutaneous island flap [5] was a suitable choice for our patient. The known advantages of this flap are an easy design with a wide range of coverage ability, a good color, texture match, and thickness of the skin, maintenance of symmetry, and balance of the lip and commissures [5]. The scars in our patient were inconspicuous due to their locations over the borders of the natural esthetic subunits and also because of the migrated new mustache that developed.

We report a rare case of SCC arising from DLE, involving the upper lip. A bilateral nasolabial orbicularis oris myocutaneous island flap is a readily available flap for total upper lip reconstruction.

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Craniometaphyseal Dysplasia

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Craniometaphyseal dysplasia (CMD) is an extremely rare genetic disease marked by progressive thickening of the craniofacial bones and aberrant development of the metaphyses in long bones. As a result of diffuse hyperostosis of the skull base, neurological symptoms

associated with cranial nerve compression, such as reduced vision, cranial nerve palsy, and deafness can occur. Craniofacial abnormalities are prominent and include hypertelorism, frontonasal bossing, a broad nasal root, prognathic mandible, and defective dentition [1]. In this paper, the authors report a case of CMD associated with facial dysmorphism and mild hearing loss in a 4-year-old girl. The clinical aspects, pathogenesis, and management of CMD will be reviewed.

A 4-year-old female was referred from the Department of Otorhinolaryngology to our department to examine her abnormal facial appearance and history of nasal obstruction and mild hearing loss. Her head circumference was in the 96th percentile, with notably wide-set eyes, a broad nasal root, a prominent forehead, and mouth breathing due to narrow nasal passages (Fig. 1). She was born normally at term with a weight of 3.1 kg. The pregnancy and neonatal period were uneventful. She had no relevant family history of skeletal or craniofacial abnormalities. The craniofacial bones were noted to have salient sclerosis and hyperostosis (Fig. 2). The distal femur was notable for a narrow diaphysis and widened metaphysis, resulting in an "Erlenmeyer flask"-shaped appearance (Fig. 3). A facial computed tomography scan exhibited reduced pneumatization of the bilateral mastoid air cells, diffuse cortical thickening of the craniofacial bones, obliteration of the paranasal sinuses, and narrowing of the cranial nerve foramina due to diffuse sclerosis of the cranial base (Fig. 4). Serum alkaline phosphatase was minimally elevated at 392 IU/L. Other characteristics were normal. The patient is cur-

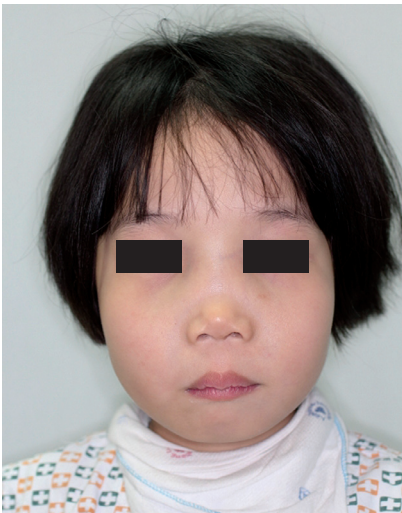


Fig. 1.
Frontal view of the patient: hypertelorism, paranasal bossing, and widening of the nasal bridge are visible.



Fig. 2.
Adenoid view of the skull shows marked sclerosis and hyperostosis of the skull base, maxilla, and mandible.