

Epidermal Nevus Syndrome with Various Skin Manifestations

— Improved with CO₂ Laser and Chemical Peeling —

Kyu Kwang Whang, M.D., Seung Min Lee*, M.D., Eun Sun Choi*, M.D., Yoon Kee Park*, M.D.

Department of Dermatology, Konkuk University College of Medicine, Chungju, Korea
*Department of Dermatology, Yonsei University College of Medicine, Seòul, Korea**

We report a case of epidermal nevus syndrome showing various skin manifestations improved with CO₂ laser and chemical peeling in a 11-year-old girl. Skin lesions were composed of linear verrucous plaques and numerous papillomatous papules on the face, neck, scalp and trunk, multiple congenital nevocellular nevi on the face, and extensive cafe au lait spots on the trunk. The associated findings of skeletal involvement were gingival hemihypertrophy and benign bone lesion of the 7th rib. CO₂ laser and chemical peeling (BCA, 50% TCA) were applied to remove these skin lesions which improved considerably. (*Ann Dermatol* 5:(1) 56-59, 1993)

Key Words: Epidermal nevus syndrome, CO₂ laser, Chemical peeling

The epidermal nevus syndrome is a congenital neuroectodermal disorder that the epidermal nevi are associated with abnormalities in other organ system¹.

Several cases have been reported and reviewed that epidermal nevi are often associated with neurologic, skeletal and other cutaneous abnormalities, and neoplasms²⁻⁷. Our case shows accompanying various types of skin lesions.

Surgical treatment has been almostly used to treat cutaneous lesions in epidermal nevus syndrome. We describe our experience with CO₂ laser vaporization and chemical peeling in treating these congenital cutaneous lesions.

REPORT OF A CASE

A 11-year-old girl visited to our department for the evaluation and treatment of multiple verrucous plaques and pigmented macules on the face, neck, scalp and trunk present since birth. None was contributory on family history.

Her general health has been good. Brown to black colored extensive multiple verrucous linear plaques were noted on the scalp, forehead, postauricular area, neck and trunk. Pinhead to rice sized black colored macules and papules were scattered on the nevus sebaceous like lesions of the face and neck, and skin colored to brown colored numerous papillomatous papules were scattered on the neck. Extensive cafe au lait spots were seen on the upper back (Fig 1, 2). Rt. gingiva were hypertrophied (Fig 3) and Rt. maxillary bone was enlarged. There was localized small bulging area on the right 7th rib which may be benign bone lesion on chest PA. Patient refused the further evaluation of this bone lesion.

The biopsy specimen taken from papillomatous lesion on the neck revealed hyperkeratosis, papillomatosis and acanthosis and that from pigmented nevus lesion showed a typical finding of nevocellular nevus (Fig. 4). The results of complete blood count, urinalysis, liver function tests, electrocardiography, electroencephalography and brain CT scan were normal or within normal limits.

Under the general anesthesia, CO₂ laser applied to linear verrucous plaque with the intensity of 2-6 watt by continuous wave mode, 2-4mm spot, and bichloroacetic acid (BCA) or 50% trichloroacetic acid (TCA) were applied to hyperpig-

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Reprint request to: Kyu Kwang Whang, M.D., Department of Dermatology, Konkuk University College of Medicine, Chungju, Korea

mented nevi after the spot test. Four weeks after treatment, the cutaneous lesions showed considerable improvement (Fig 5).



Fig. 1. Facial lesions composed of epidermal nevi and numerous congenital nevocellular nevi.



Fig. 3. Rt. gingival hypertrophy with verrucous mucosal changes.



Fig. 2. Picture of the back showing extensive cafe au lait spots, nevocellular nevi and epidermal nevi.

DISCUSSION

The epidermal nevi are organoid nevi arising from pluripotential germinative cells of the basal layer of the embryonic epidermis. Epidermal nevus syndrome can be described when epidermal nevi are associated with congenital skeletal, central nervous system abnormalities¹. Epidermal nevi are at significant risk of having other organic abnormalities and warrant detailed initial assessment and close follow up. Appropriate procedures and laboratory studies might be required when there are the abnormal findings on the history and physical examination.

Various cutaneous associations of epidermal nevi have been described. Solomon and Esterly⁸ reported the associations of various vascular nevi, areas of hypopigmentation, large and small cafe au lait spots, multiple acquired nevocellular nevi, small congenital nevocellular nevi and dermatom-

galy in their cases.

Our patient manifested various cutaneous abnormalities of the epidermal nevus syndrome. She had many epidermal nevi with nevus sebaceous like lesions on the scalp and the face, congenital nevocellular nevi on the face, nevus lateris on the



Fig. 4. a) Verrucous epidermal nevi before treatment.
b) After 4 weeks of CO₂ laser vaporization.

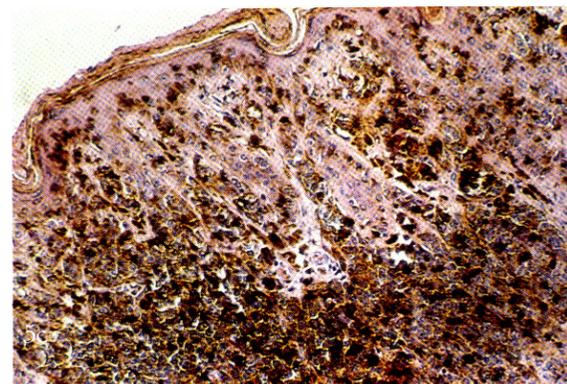
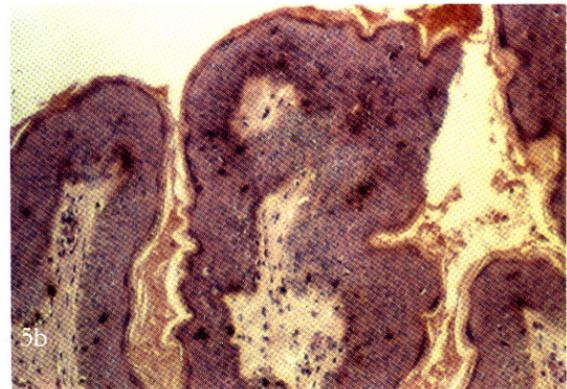


Fig. 5. Histopathologic findings.
a) Epidermal nevi lesions show hyperkeratosis, papillomatosis, acanthosis and hyperpigmentation of the basal cell layer (H-E stain, $\times 100$).
b) Papillomatous changes on the posterior neck (H-E stain, $\times 40$).
c) Typical histopathologic findings of congenital nevocellular nevi (H-E stain, $\times 200$).

trunk and extremities, large cafe au lait spots on the trunk, and numerous small papillomas on the neck.

It was reported that the concomitant involvements of oral cavity in the epidermal nevus syndrome was extension of nevi to the upper lip and hard palate, cleft palate, bifid uvula, hypoplastic teeth, odontomas and odontogenic tumors such as ameloblastoma^{9, 10}. Our patient had skeletal abnormalities like Rt. gingival hemihypertrophy and benign bone lesion on the 7th rib.

Surgical excision is the most effective treatment of epidermal nevi. Excision may be impractical when nevi are extensive. For large nevi, a step-wise excision is recommended. When it is not advisable, several alternative treatment methods have been reported using liquid nitrogen, cryotherapy, podophyllin, retinoic acid and dermabrasion¹¹⁻¹³. Other modalities such as topical coal tar with or without ultraviolet light, radiation therapy, anthralin, and 5-Fluorouracil have been tried without success¹¹.

The CO₂ laser has been applied frequently in the dermatology and dermatologic surgery fields¹⁴⁻¹⁶. It is versatile, being either a vaporizing tool for proliferative disorder or a light scalpel. The postoperative period is relatively painless for the patient. When energy from the carbon dioxide laser beam strikes soft tissue, there is instantaneous conversion of cellular water into steam and smoke at 100°C. Ratz et al¹⁶ reported their successful experience with CO₂ laser vaporization in the treatment of epidermal nevi.

Chemical peeling is a relatively common dermatologic procedure. The depth and effectiveness of the chemexfoliation obtained will vary with the peeling agent. Trichloroacetic acid (TCA) is now commonly utilized as a peeling agent for treatment of aging, wrinkled, and sun damaged skin¹⁷. Rosian et al¹⁸ used bichloracetic acid (BCA) in the treatment of benign sebaceous hyperplasia.

We applied the CO₂ laser to epidermal nevi lesions on the scalp and forehead, and applied the BCA and 50% TCA to nevocellular nevi on the face under the general anesthesia. The postoperative period was painless. At the postoperative period 4 weeks, epidermal nevi lesions showed marked flattening and congenital nevocellular nevi

lesions showed lessening pigmentation.

Either the CO₂ laser or chemical peeling can be a kind of modality to help removing various cutaneous lesions in epidermal nevus syndrome.

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