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Carbon Dioxide-Laser Treatment of Trichoepitheliomas in Brooke-Spiegler Syndrome

Síndrome de Brooke-Spiegler: tratamiento de los tricoepiteliomas con láser de CO_2

To the Editor:

Brooke-Spiegler syndrome is a rare genodermatosis that causes a predisposition to adnexal tumors due to alterations in the folliculosebaceous apocrine unit.[1-4] It is an autosomal dominant condition with variable penetrance and is characterized by the simultaneous and progressive appearance of multiple cylindromas on the scalp, facial trichoepitheliomas, and, occasionally, eccrine spiradenomas.[1-4]

The locus implicated in this condition is found on chromosome 16q12-q13, a region that holds the *CYLD1* tumor suppressor gene, which is involved in the regulation of proliferation of the skin appendages.[5] There is marked phenotypic variability within families, such that members of a single family with identical mutations can present isolated or multiple trichoepitheliomas, cylindromas or, less frequently, eccrine spiradenoma.

Trichoepitheliomas present clinically as small, translucent papular lesions that are painless and are most commonly found in groups in the nasolabial sulci, on the nose, and on the forehead. Histologically there are multiple islets of basaloid cells in a fibrous stroma associated with numerous corneal cysts. There have been very rare reports of malignant transformation in multiple familial trichoepitheliomas and of the associated presence of basal cell carcinomas.[6] The differential diagnosis includes hereditary disorders that present with multiple firm facial papules such as Birt-Hogg-Dubé syndrome (fibrofolliculomas), Cowden syndrome (tricholemmomas), tuberous sclerosis (angiofibromas), multiple basaloid follicular hamartoma syndrome (basaloid follicular hamartomas), Rombo syndrome (trichoepitheliomas), and Gardner syndrome (epidermal cysts).[7]

Cylindromas appear as multiple, well-defined, firm pink nodules with a smooth surface and superficial telangiectasias, and a diameter that varies from a few millimeters to several centimeters; they are found on the head, particularly on the scalp, and may occasionally be painful. Histologically, the lesions present as well-defined dermal nodules not in contact with the epidermis; they are formed of dense islets of basaloid cells arranged in a jigsaw-puzzle pattern and surrounded by a highly eosinophilic material.

Both trichoepitheliomas and cylindromas tend to increase in size and number over time and can lead to pronounced cosmetic alterations with psychological, social, and occupational repercussions. Early treatment is therefore indicated in order to reduce postsurgical sequelae and increase patient satisfaction.

Various palliative treatment approaches have been described in the literature, including electrocoagulation, cryotherapy, dermabrasion, tricholoracetic acid, retinoic acid, carbon dioxide (CO_2) laser, radiation therapy, and surgery.[8,9]

We present 2 cases of multiple trichoepitheliomas treated using CO_2 laser. The first patient was a 43-year-old woman with Brooke-Spiegler syndrome. Over 9 years she had received 4 sessions of continuous-wave CO_2 laser vaporization (sessions every 2 years) with a power setting of 3 to 5 W and 1 to 3 passes (Figures 1 and 2). The second patient was a 43-year-old man with Brooke-Spiegler syndrome. Since 2003 he had undergone sessions every 2 years of continuous-wave CO_2 laser vaporization using a power of 5 W (Figure 3).

In both cases the cosmetic result had been satisfactory, achieving a significant reduction (flattening) of the lesions, though they had never disappeared completely. Over time there had been a gradual recurrence, associated with the appearance of new lesions, but there was an adequate response to further CO_2-laser treatments.

The CO_2 laser is a surgical instrument that emits energy in the form of infrared light at a wavelength of 10 600 nm; this wavelength is absorbed by water, leading to vaporization of the skin with coagulative necrosis in the remaining dermis. Used in continuous mode it produces a surgical cut (focused beam) or vaporization (defocused beam). The most important characteristics of this laser are its rapid action, permitting large areas to be treated, the selectivity and precision of its effects, and its high specificity of tissue damage, producing highly localized destruction and enabling multiple lesions to be treated with minimal bleeding. The complications of treatment include erythema, edema, a burning sensation, Herpes simplex

Figure 1 A 43-year-old woman with Brooke-Spiegler syndrome. Treatment by continuous-wave carbon dioxide laser vaporization: before (left) and 5 months after (right) treatment.

Figure 2 Same patient as in Figure 1. The trichoepitheliomas shown in greater detail before (left) and after (right) treatment.

Figure 3 A 43-year-old man with Brooke-Spiegler syndrome (left). Scab formation immediately after vaporization by continuous-wave carbon dioxide laser at a power of 5 W (right).

reactivation, hypopigmentation or hyperpigmentation, and hypertrophic scars.[10] It is used particularly in rhinophyma, actinic cheilitis, Bowen disease, trichoepitheliomas, neurofibromas, syringomas, palpebral xanthelasma, facial angiofibromas, lymphangioma circumscriptum, angiokeratoma, epidermal nevus, congenital nevus, and Hailey-Hailey disease. It can also be used for warts, keloids and hypertrophic scars, actinic keratoses, basal cell carcinoma, and radiation dermatitis.[8]

Carbon dioxide laser therapy, ever more widely used in dermatology, was a good therapeutic option in the cases of Brooke-Spiegler syndrome described here, as the large number of facial lesions can often produce a significant psychological impact and we were able offer a treatment that, although not curative, achieved good cosmetic results with minimal side effects.

Conflict of Interest

The authors declare that they have no conflict of interest.

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