Pseudoxanthoma Elasticum Treatment with Fractional CO₂ Laser

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Summary: Pseudoxanthoma elasticum (PE) is a rare genetic disease characterized by calcification and fragmentation of elastic fibers of the skin, retina, and cardiovascular system. We report a case of PE in which fractional carbon dioxide laser treatment was successfully used to achieve improvement of the cervical skin with 2-year follow-up, in a patient with Fitzpatrick skin type IV. After the fifth session, the patient presented with a local herpes infection. The postlaser reaction of the PE skin was similar to that of the normal skin, in terms of the duration of redness, pain, swelling, and duration of crusting. The overall cosmetic result was satisfactory, with improvement in skin texture, irregularity, volume, and distensibility. The herpetic infection reinforces the value of antiviral prophylaxis during laser treatment of extrafacial areas. (Plast Reconstr Surg Glob Open 2014;2:e219; doi: 10.1097/GOX.0000000000000173; Published online 26 September 2014.)

Pseudoxanthoma elasticum (PE) is a rare autosomal recessive disease caused by mutations in ABCC6 (adenosine triphosphate-binding cassette transporter C6), located in chromosome 16. Its prevalence is estimated to be 1:50,000. PE involves progressive calcification and fragmentation of the elastic fibers in the skin, retina, and cardiovascular system. The initial symptoms can occur at any age and are usually cutaneous: painless dyschromias or yellow-orange papules, commonly in the lateral and posterior cervical regions and in the axillary, inguinal, popliteal, and periumbilical areas. The most frequent cause of morbidity is reduction of visual acuity. The diagnosis is based on clinical aspects, as the lesions are very unique, and on genetic study and histological examination. Several treatments have been described for the dermatological lesions of PE: surgical excision, collagen or autologous fat injections, and even therapeutic abstention. However, none of these treatments is completely effective or definitive.

Carbon dioxide (CO₂) laser skin resurfacing has been a mainstay of facial rejuvenation since its introduction in the mid-1990s. Recently, fractional CO₂ lasers were proven safe and effective not only for facial rejuvenation but also for treatments elsewhere on the body. Because of the areas of spared epidermis and dermis, these lasers are associated with more rapid healing than fully ablative CO₂ laser skin resurfacing, and downtime (recuperative period after a treatment) is proportionately reduced.

The objective of this study is to present a case of PE treated with fractional CO₂ lasers.

CASE REPORT

Our patient was a 32-year-old woman, with Fitzpatrick skin type IV, who was diagnosed with PE 17 years ago. She had cutaneous involvement of the cervical, axillary, and periumbilical regions and a history of labial herpes. Her brother also had PE with cervical...
involvement. The patient signed an informed consent.

The initial treatment was with 0.025% tretinoin cream and sunscreen. After 3 months, treatment with a fractional CO₂ laser (SmartXide Dot; Deka, Florence, Italy) was applied only in the posterior cervical region (30 W; spacing 750; dwell time 500). At the second session, treatment of the anterior cervical region was included. She underwent further treatments at 2-month intervals, and the laser parameters were adjusted according to the previous treatment outcomes (Table 1). In total, she had 4 sessions for the anterior cervical region and 5 sessions for the posterior cervical region. Dipyrone 1.0 g and cold air were administered to decrease pain (Freddo; Fabinject, Taubaté, Sao Paulo, Brazil). The patient assessed pain sensations after each session on a scale from 0 to 10. The laser and clinical parameters for each session, including duration of pain, edema, hyperemia, and scabs, are shown in Table 1.

At 15 days after the last session, she developed watery blisters or vesicles, with discharge and itching, in the cervical region (Fig. 1). The results of a biopsy were consistent with herpes simplex, and she was treated with acyclovir (1 g/d for 20 days), cephalexin, and topical betamethasone in the first 6 days. Complete resolution of the symptoms occurred after 20 days, but this area showed postinflammatory hyperpigmentation, which persisted for 18 months, when treatment with 0.015% tretinoin cream and 2% hydroquinone was prescribed with partial improvement.

The treatment outcome was evaluated using a scar scale, which was composed of 5 parameters\(^{14}\) and was filled separately by both the patient and the medical staff (average of 2 independent medical evaluators) (Tables 2 and 3). The evaluations were performed between each session and at 3, 6, 12, and 24 months after the last session.

A significant improvement in skin irregularity and elasticity was already evident 1 month after the first session. At 2 years after the last treatment, there was improvement of surface irregularities and distensibility in the cervical area. Both the patient and the medical staff were satisfied with the results (Figs. 2, 3).

### Table 1. Laser Parameters and Clinical Progress After Each Fractional CO₂ Laser Session

| Session No. | Spacing | Dwell Time | Energy/Point (mJ) | Fluency (J/cm²) | Density of Ablation | Pain Intensity 0–10 | Pain (hr) | Edema (d) | Crusts (d) | Hyperemia (d) |
|-------------|---------|------------|-------------------|-----------------|---------------------|---------------------|----------|-----------|-----------|-------------|
| I           | 750     | 500        | 15                | 1.24            | 8.0%                | 9                   | 24       | 2         | 3         | 2           |
| II          | 500     | 800        | 24                | 3.32            | 13.3%               | 6                   | 5        | 2         | 5         | 2           |
| III         | 500     | 1000       | 30                | 4.15            | 13.3%               | 6                   | 3        | 2         | 8         | 2           |
| IV          | 500     | 1200       | 36                | 4.98            | 13.3%               | 6                   | 2        | 1         | 7         | 1           |
| V           | 300     | 1200       | 36                | 8.52            | 22.8%               | 9                   | 72       | 2         | 5         | 2           |

Pain was subjectively assessed by the patient immediately after each session.

### Table 2. Evaluation of Treatment Outcome by the Patient

| Description     | Color | Texture/Hydration | Surface Irregularities | Volume | Distensibility | Total | Subjective Score (0–10) |
|-----------------|-------|------------------|------------------------|--------|----------------|-------|------------------------|
| Prelaser I      | 1     | 1                | 0                      | 0      | 0              | 2     | 7                      |
| Prelaser II     | 1     | 1                | 0                      | 0      | 1              | 4     | 8                      |
| Prelaser III    | 2     | 1                | 1                      | 1      | 1              | 6     | 8                      |
| Prelaser IV     | 2     | 1                | 1                      | 1      | 1              | 6     | 8                      |
| Prelaser V      | 2     | 1                | 1                      | 1      | 1              | 6     | 9                      |
| 3 mo postlaser  | 1     | 2                | 1                      | 1      | 1              | 6     | 9                      |
| 6 mo postlaser  | 2     | 2                | 2                      | 1      | 2              | 9     | 9                      |
| 1 yr postlaser  | 1     | 1                | 1                      | 1      | 1              | 6     | 8                      |
| 2 yr postlaser  | 1     | 1                | 1                      | 1      | 2              | 6     | 7                      |

Color, texture/hydration, surface irregularities, volume, and distensibility were evaluated on the following scale: 0 = bad, 1 = regular, and 2 = good. “Total” indicates the sum of the previous 5 columns. The last column shows the subjective score, from 0 to 10, for comparison with the score obtained using the scar scale.
Lasers can be used to treat a variety of medical dermatological conditions, such as acne vulgaris, psoriasis, and vitiligo. Fractional CO$_2$ laser ablation improves the appearance of wrinkles, photoaging, sagging skin, acne scars, and burns, without the disadvantages and risks related to prolonged epithelialization after traditional ablative lasers, such as prolonged erythema, hypopigmentation, and even scarring.

In the present case, a fractional CO$_2$ laser was used for the treatment of cutaneous PE lesions. This treatment has not been previously used to treat this pathology. We began laser treatment cautiously, using low energy, and then progressively adjusted

Table 3. Evaluation of Treatment Outcome by the Doctor

| Description | Color | Texture/Hydration | Surface Irregularities | Volume | Distensibility | Total |
|-------------|-------|-------------------|------------------------|--------|---------------|-------|
| Prelaser I  | 1     | 0.5               | 0                      | 0      | 1             | 2.5   |
| Prelaser II | 2     | 1.5               | 1                      | 0      | 1             | 5.5   |
| Prelaser III| 2     | 1                 | 1                      | 1      | 1             | 6.0   |
| Prelaser IV | 2     | 1.5               | 1                      | 1      | 1             | 6.5   |
| Prelaser V  | 1     | 1.5               | 1                      | 1      | 1             | 5.5   |
| 3 mo postlaser | 1 | 1.5              | 1                      | 1      | 1.5           | 6.0   |
| 6 mo postlaser | 1 | 1.5              | 1                      | 1      | 1.5           | 6.0   |
| 1 yr postlaser | 1  | 1.5              | 1                      | 1      | 1.5           | 6.0   |
| 2 yr postlaser | 1  | 1                | 1                      | 1      | 2             | 6.0   |

Color, texture/hydration, surface irregularities, volume, and distensibility were evaluated on the following scale: 0 = bad, 1 = regular, and 2 = good. “Total” indicates the sum of the previous 5 columns.

Fig. 2. Before (A) and 2 years after (B) treatment, frontal view.

Fig. 3. Before (A) and 2 years after (B) treatment, side view.
the treatment due to the good outcomes. After the laser treatment, the reaction of the PE-affected skin was similar to that of the normal skin, in terms of erythema, pain, swelling, and duration of crusting. With increase in the dwell time, there was a tendency for longer duration of crusts.

Despite the reactivation of herpes after the last session, in which the density of the treatment was increased, there was a slow but satisfactory resolution of the PE symptoms, and fortunately, the end result was not affected. This observation highlights the importance of antiviral prophylaxis in patients with a history of herpes, even when the laser treatment is extrafacial.

Our patient’s clinical progress was favorable and satisfactory. The scar scale showed good sensitivity in the assessment of skin improvement. The final result after 2 years was an improvement in skin texture, surface irregularities, and distensibility.

CONCLUSIONS
We have reported the first instance of fractional CO₂ laser treatment of cutaneous PE lesions. This treatment produced appropriate healing and good esthetic results.

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