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Case Reports

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Treatment of neurofibromatosis NF-1 with CO₂ laser - Case report

Tratamento de neurofibromatose NF-1 com laser de CO₂ - Relato de caso

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ABSTRACT

Type 1 neurofibromatosis has multiple cutaneous lesions and limited treatment options. CO₂ laser is a useful tool for the removal of neurofibromas, the main source of cosmetic disfiguration for these patients. We present the treatment of neurofibromas with CO₂ laser in a patient over 4.5 years. The patient had lesions with variable diameter, diffusely distributed. After 16 sessions of CO₂ laser, we observed clinical improvement. Although it is a palliative treatment, the cosmetic improvement achieved substantiates the demanded effort and time in view of the psychological gains for the patient.

Keywords: *Lasers, Gas; Laser Therapy; Neurofibromatosis 1*

RESUMO

Introdução: A neurofibromatose tipo 1 apresenta lesões cutâneas múltiplas e limitadas opções terapêuticas. O laser de CO₂ é ferramenta útil na remoção de neurofibromas, a principal fonte de desfiguração cosmética nesses pacientes. Apresentamos o tratamento de neurofibromas com laser de CO₂ em uma paciente ao longo de quatro anos e meio.

Relato do caso: A paciente apresentava lesões de diâmetro variável, distribuídas difusamente. Após 16 sessões com laser de CO₂ constatou-se melhora clínica.

Conclusão: Apesar de ser tratamento paliativo, a melhora estética obtida justifica o esforço e tempo demandados, tendo em vista os ganhos na esfera psicológica do paciente.

Palavras-chave: *Lasers de Gás; Neurofibromatose 1; Terapia a Laser*

INTRODUCTION

Neurofibromatosis type I (NF1), previously known as Von Recklinghausen disease, is a dominant autosomal condition with variable incidence (1:2,500 to 1: 3,000 inhabitants). It results from loss of expression of the NF1 gene, which is responsible for the synthesis of neurofibromin, a protein that plays a role in the mechanisms that regulate cell proliferation. It is observed in all races and is unrelated to gender. Neurofibromatosis type I may involve changes (neurofibromas and cutaneous hyperpigmentation) in multiple systems, including the nervous system, bones and skin. Neurofibromas are benign peripheral nerve tumors that can be observed in 48% of patients at 10 years of age.¹⁻²

The National Institutes of Health (NIH) has established a consensus on the diagnostic criteria for NF1. Two or more of the following findings are needed: six or more *café-au-lait* patches; two or more cutaneous / subcutaneous neurofibromas or one plexiform neurofibroma; axillary or inguinal ephelides;

optic glioma; two or more Lisch nodules in the eyes; dysplasia of the sphenoid bone or cortical thinning of long bones and a first degree relative with NF1. Any region of the body can be affected by cutaneous neurofibromas, which can vary in size, number and distribution. Usually they appear at puberty and continue to grow. Deformities can result from the growth of hundreds of cutaneous neurofibromas, leading to social isolation and emotional distress.²

The treatment of this cutaneous alteration is predominantly surgical, however alternative treatments should be considered for patients with multiple lesions – usually above 100 – in whom surgical intervention is not possible or desirable. CO₂ laser has been shown to be effective in the treatment of large to medium neurofibromas in large numbers, with similar or better aesthetic outcomes than those obtained with surgical excision. However, evidence of CO₂ effectiveness, treatment effects on patients' satisfaction, and post-treatment recurrence rate are scarce.³⁻⁵

The present paper describes the case of a 37-year-old patient bearing hundreds of dermatological lesions of neurofibromatosis type I, treated with CO₂ laser for four and a half years, as well as the results achieved.

CASE REPORT

A 37-year-old, single female patient originary from the Brazilian Northeast City of Aracaju (SE) referred dark spots on the body (six in number), from birth. At 23 years of age, she noticed the onset of lesions distributed throughout the integument. She denied family history of this pathology.

Physical examination allowed the observation of numerous papular and nodular cutaneous lesions, with diameters varying from 1.0 to 15.0 mm throughout the integument, mainly the face, neck, cervix and abdomen, and in smaller number in the limbs and dorsum. There was presence of brownish *café-au-lait* patches on the abdomen, chest, back and lower limbs.

Treatment started at the age of 26, when the patient underwent 23 surgical excisions, having had another 16 lesions removed later on. At the age of 37, 30 additional lesions were resected.

Treatment of lesions with CO₂ laser began at 37 years of age.

Procedure

Firstly, a test procedure was performed on three submandibular lesions. Two months after, good healing was observed, with absence of interurrences and remission of the treated lesions. Thus, the treatment of the remaining lesions was started.

The Laser Pixel CO₂ 70W, Alma Lasers (Halamish St. Caesarea Industrial Park, Israel) was used with the surgical tip, 2.5w, 10ms on-time, 20ms off-time. Lesions up to 3 mm in size were treated with the handpiece positioned perpendicularly to the surface, with circular movements. Larger lesions were initially cauterized at the periphery and then clamped at the base, with forced extrusion aiming at facilitating their delimitation. Surgical sutures were performed only in lesions larger than 1.0



FIGURE 1: Pre and post-treatment (six sessions)

cm. Post-cauterization curettage of lesions was carried out in the first sessions in order to confirm the complete ablation of the neurofibromas. On average, 50 to 100 lesions were treated per session, depending on their size. (Figure 1)

Sixteen sessions were carried out over four and a half years, with intervals varying from two weeks to ten months. A decision was made for treating a specific region's lesions per session, prioritizing the body sites of greater visibility (face, neck, neck, forearms, dorsum of the hands), and at a later stage later the chest, abdomen, arms and back. (Figures 2, 3 and 4)

Clinical development

Healing by second intention occurred in two or three weeks. There was no recurrence of the facial lesions; nevertheless there were recurrence of several lesions in the thorax, as well as in the abdominal region due to incomplete cauterization. The scars on the face and thorax remained erythematous for one to four months, developing into hypopigmentation and sometimes into atrophy. There were no additional complications.

DISCUSSION

The standard procedure for the removal of NF1 lesions is surgical excision, which has the advantage of leading to a linear scar. Disadvantages are longer procedure durations and greater risk of bleeding.³⁻⁴ In light of this, CO₂ laser treatment is more suitable for the removal of large numbers of lesions with reasonable aesthetic outcomes and low risk of complications.⁶⁻⁷

Previous studies have shown that CO₂ laser treatment has improved the patients' self-confidence as well as their social and sexual lives. Algermissen *et al.*⁴ reported that depigmented, laser treated scars were more acceptable to patients than scars resulting from surgical excisions, a finding that is in line with the opinion of the patients in the present study.

Table 1 shows a summary of the previously reported physical and psychological benefits of using CO₂ laser treatment.

The patient treated by the authors of the present report showed a high level of satisfaction, with hypopigmentation at



FIGURA 2: Pré-tratamento e pós-tratamento com seis sessões



FIGURA 3: Pós-procedimento imediato de face e colo



FIGURA 4: Pré-tratamento e pós-tratamento com três sessões

TABLE 1: Summary of physical and psychological benefits

Studies	Main findings
Moreno <i>et al.</i> ⁵	82% reported a good level of improvement in symptoms (e.g. pain, pruritus) and social activity 73% reported a good level of improvement in sexual activity
Algermissen <i>et al.</i> ⁴	100% reported a good level of improvement in the feeling of <i>despair</i> satisfaction and recommendation rate = 73%
Chiang <i>et al.</i>	Most patients reported increased self-confidence and social acceptance Satisfaction rate = 92% Recommendation rate = 100%

the treatment sites being the only adverse effect described, suggesting that the treatment with CO₂ laser has great potential in the improvement of the psychological well-being of patients suffering from this pathology. To date, no previous reports with long-term follow-up have been published, as was the case with the present paper.

CONCLUSION

Although there is no effective treatment to revert lesions characteristic of NF1, CO₂ laser is an option associated to a high level of patient satisfaction and swiftness in the approach of lesions, allowing to treat a large number of lesions per session, with a low risk of complications. ●

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