Multiple facial angiofibromas treated with high-frequency equipment

Tratamento dos angiofibromas múltiplos da face com radiofrequência

Allysson Antonio Ribeiro Gomes
Ylka Virginia Ribeiro Gomes
Felipe Barbosa Lima
Salustiano Gomes de Pinho Pessoa

Resumo: Esclerose tuberosa é uma doença genética rara, com herança autossômica dominante, associada à formação de hamartomas múltiplos em vários órgãos, como cérebro, pele, pulmões, rins, coração e olhos. Os autores deste estudo apresentam um caso de uma paciente do sexo feminino, com 30 anos de idade, portadora de esclerose tuberosa, apresentando múltiplos angiofibromas em face, tratada com equipamento de alta frequência (radiofrequência), e discutem as opções terapêuticas para tratamento de indivíduos portadores de esclerose tuberosa com extenso envolvimento cutâneo.

Palavras-chave: Angiofibroma, Eletrocirurgia; Esclerose tuberosa

Abstract: Tuberous sclerosis is a rare genetic disease with autosomal dominant inheritance, associated with multiple hamartomas in several organs, such as the brain, skin, lung, kidney, heart and eyes. The authors of this study report a case of a 30 years old female patient with tuberous sclerosis, presenting multiple angiofibromas on face treated with high frequency equipment (radiofrequency), and discuss the therapeutic options for treatment of individuals with extensive cutaneous involvement in tuberous sclerosis.

Keywords: Angiofibroma; Electrosurgery; Tuberous sclerosis

INTRODUCTION

Tuberous sclerosis (TS) is an autosomal dominant neurocutaneous geno-dermatosis characterized by the formation of hamartomas in multiple organs: brain, skin, lungs, kidneys, heart and eyes. It affects approximately 1 in every 6,000 to 10,000 individuals. The skin is involved in around 90% of the cases, and the signs associated with TS are hypomelanic macules, angiofibromas, fibrotic plaques on the forehead, periungual fibromas, cutaneous plaques with “orange peel” aspect, gingival fibromas, and “café-au-lait” macules. During puberty the angiofibromas proliferate to involve the nose and cheeks, symmetrically, becoming a source of great psycho-social and hygiene issues. The treatment of these lesions varies from removal with surgical blade to dermabrasion, CO2 laser, argon laser and radiofrequency ablation. The present case study aims to report a case of surgical treatment using radiofrequency ablation and to revise the therapeutic options for angiofibromas on the face in patients with TS.

CASE REPORT

Female patient, 30 years old, seen at the Plastic Surgery unit at Hospital X, sent to the dermatology service for treatment of disfiguring angiofibromas on the face (Figure 1). The initial diagnosis of TS was con-
firmed by the neurologic service using the criteria table. The clinical history included convulsive fits since the age of eight months associated to cognitive deficit, without ocular lesions. The skin involvement started at the age of five, with hypolelanic macules on the back, fibrotic plaques on the forehead and cutaneous plaques with “orange peel” aspect on the lumbar region. The facial involvement started on the dorsum of the nose when she was eight years old, and there was a report of previous treatment by shaving at the age of 13, without success. The family members complained of deformity and distortion of the aesthetic units of the face, which prevented social and school life, leading to great psycho-social discomfort.

At physical examination the lesions consisted of reddish-brown, vegetative, shiny, elastic tumors, all over the nose, partially obliterating the nostrils, malar regions, lower lip, chin, and, in smaller numbers and non-pedunculated, on the zygomatic areas and forehead. Also on the forehead there were fibrotic plaques measuring 0,5 to 2,5 cm. On the lids, as well as on the neck and trunk the lesions were pedunculated, similar to syringomas. The lesions had a fetid smell due to the accumulation of sebaceous secretion and sweat, associated to the difficulty in relation to local hygiene. Also noted were spontaneous peringual fibromas and cutaneous plaques with “orange peel” aspect.

The biopsy revealed dense fibrous tissue with vascular proliferation with no evidence of malignity, and the clinical and histopathological diagnosis was angiofibroma.

Surgical technique: under general anaesthesia the first surgical stage was performed with the removal of the lesions on the nose, malar regions bilaterally and upper lip. During the second surgical stage the lesions on the chin, mandible and lower lip were removed, and the lesions on the upper and lower lids were resected.

High frequency device was used for the procedure. The chosen electrode was the loop, 0,5 cm thick and 1,2 cm in diameter. Chlorhexidin was used for asepsis and antisepsis and the surface was kept moist.

The superficial layers were roughly resected, followed by more delicate movements, no deeper than the deep dermis, to delineate the contours of the dorsum of the nose, alar lobules and nostrils. During the same procedure the electrocautery, dermabrasion with high rotation engine with diamond tips, and cold blade were tested. All methods were effective on the removal of the lesions, however they caused more bleeding. At the end, the whole area was cleaned with 0.9% saline solution and protected with open dressing with cicatrizing ointment.

RESULTS

The patient remained 24 hours under observation in the hospital. She was asymptomatic and was released with orientation in regards to local care and dressings and, after healing, use of sunscreen. Subsequent evaluations were performed on the 7th, 14th and 30th postoperative days, and then monthly for the first four months. After five months of follow-up there were no signs of scars or cutaneous despigmentation. After the third postoperative month recurrence of some lesions in smaller size and number was noted. The treatment was successful in getting the patient back to social life and a great improvement was noted in regards to cosmetic aspect and hygiene (Figure 2).

DISCUSSION

Tuberous sclerosis is a rare genetic disease caused by the mutation of two genes: TSC 1 on chromosome 9q34 and TSC2 on chromosome 16p13.3. The pathogenesis results from the expression and action of the products of these genes: the proteins tuberin and hamartin, respectively, in all tissues that, simultaneously, act on the modulation of cellular growth. The disease affects primarily the skin and the
central nervous system but tumors can also develop on the kidneys, eyes, heart and lungs. Around 96% of the patients have at least one cutaneous presentation. The angiofibromas start to appear between the 1st and 4th years of age and typically grow during puberty, when they can be mistaken by acne vulgaris. The differential diagnosis also includes the multiple papulo-nodular lesions of the basal cell nevus syndrome, verruca plana, syringomas, benign pedunculated tumors and sarcoidosis. When they are extensive and typically bilateral they are pathognomonic of TS, and a biopsy should be conducted in order to ascertain the diagnosis. Serious psycho-social disturbances affect specifically teenagers with mild cognitive deficit and cosmetic treatment of the lesions is recommended.

Literature review reports multiple treatment modalities for the facial angiofibromas. Liquid nitrogen has been used with good results. Electrosurgery followed by electrocauterization has been tried but a high risk of pathological scars and cutaneous pigmen-
tary alterations was observed. We noted increased bleeding during the surgical process with this technique which led to it being abandoned. Chemical peels and oral use of acid 13-cis-retinoic were tested by the same author with variable results. Various types of laser have been tried: argo laser, CO2 abrasion, copper vapour, and potassium titanyl phosphate laser. According to Verma, lasers are recommended as the treatment of choice for early lesions and small vascular ectasies, with low level of recurrence and low incidence of scars. The same author reports an improvement of 50 to 80% in patients with extensive lesions using CO2 laser with irradiation of 320 watts/cm² up to 63,000 watts/cm², keeping the device at variable distances from the skin to be treated. According to El Musa ET AL, the negative aspects of the use of lasers are the need for multiple sessions for the same areas, long operatory time specially with the argon laser, which penetrates and vaporizes only a few centimetres of tissue, and the very high costs involved with infra structure, equipment and training. With the use of radiofrequency and not going beyond the deep dermis we obtained 90% of improvement on the treated area in one single session. Other authors advocate the use of dermabrasion associated with shaving as a satisfactory method, with low recurrence of the lesions and the possibility of treating the whole face in a single procedure, with reduced surgical time and low costs. However we observed with this method the dissemination of particles in aerosol and important bleeding when treating hypervascular lesions.

The use of high frequency for the ablative treatment of angiofibromas was firstly described by Swarrop ET AL in 2008. Radiosurgery (electrosurgery or high frequency surgery) is the process of cutting and/or coagulation of tissues using a high frequency alternate electric current (frequency used in FM radio) and low amperage. The energy generated by the passing of the radiofrequency waves causes the cytoplasm to boil, rupturing the cell form inside out, a phenomenon called cell volatilization. The present radiosurgery equipments, as well as generating relatively pure waves of cutting and coagulation, also produce blended waves which allows for excellent cutting and haemostasis. It also enables the amount of haemostasis not to be affected by the potency adjustment of the cutting module. There is also minimum lateral spreading of the energy, propitiating a better cosmetic result and easier delimitation of the treated area. We observed minimum bleeding and it was easy to delineate the anatomic contour of the nostrils, as described by Sperli for the treatment of rhinophyma. Radiofrequency is a safe, effective and economic treatment option for extensive angiofibroma on the face, however recurrence may occur and new interventions might be necessary.

FIGURE 2: Postoperatory: A= frontal vision; B= nasal pyramid
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MAILING ADDRESS / ENDEREÇO PARA CORRESPONDÊNCIA:
Allysson Antonio Ribeiro Gomes
Rua Coronel Américo Porto, 303 - apartamento 701, edificio Porto Seguro, Lauritzen
CEP 59401-381 Campina Grande (PB) - Brazil
e-mail: dr.allyssongomes@hotmail.com

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