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Idiopathic harlequin syndrome: successful therapy with botulium toxin

Síndrome de arlequim idiopática: sucesso terapêutico com toxina botulínica

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ABSTRACT

Harlequin syndrome is a rare autonomic disorder characterized by anhidrosis and lack of unilateral flushing of the face, which may affect the cervical and thoracic regions. Paradoxically, there is compensatory flushing and sweating on the contralateral side to the alteration. It is idiopathic in most cases, but it can be congenital or secondary to structural or post-surgical iatrogenic lesions. Treatment is directed at the causative factor. We describe the case of a patient with a diagnosis of idiopathic Harlequin Syndrome with botulinum toxin application in the hemiface affected by compensatory symptoms with good therapeutic response.

Keywords: Hyperhidrosis; Blushing; Botulinum toxin type A

RESUMO

A síndrome de arlequim é uma rara desordem autonômica que se caracteriza por anidrose e falta de rubor unilateral da face, podendo acometer as regiões cervical e torácica. De forma paradoxal, há rubor e sudorese compensatórios no lado contralateral à alteração. É idiopática na maioria dos casos, mas pode ser congênita, secundária a lesões estruturais e à iatrogenia pós-cirúrgica. O tratamento é direcionado ao fator causal. Descreve-se caso de paciente com diagnóstico de síndrome de arlequim idiopática, sendo realizada aplicação de toxina botulínica na hemiface acometida pelos sintomas compensatórios com boa resposta terapêutica.

Palavras-chave: *Hiperidrose; Rubor; Toxinas botulínicas tipo A*

Letters

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INTRODUCTION

Harlequin syndrome is a rare autonomic disorder characterized by anhidrosis and a lack of unilateral facial flushing, which can affect the cervical and thoracic regions.[1] Paradoxically, there is compensatory flushing and sweating on the side contralateral to the alteration.

CASE REPORT

A 35-year-old woman reported erythema and sweating on the right hemiface for two years, on exertion and heat.

Upon dermatological examination, after performing physical exercise for 20 minutes, she presented erythema with a well-defined demarcation line on the hemiface, cervical region, and upper chest on the right, associated with intense ipsilateral sweating. There was no pupillary alteration or palpebral ptosis (Figures 1A and 3A).

Imaging tests excluded compressive causes. Magnetic resonance imaging of the skull, neck, and cervical spine and chest tomography showed no alterations. Neurological and ophthalmological evaluations showed no noteworthy findings. Thus, we complete the diagnosis of idiopathic harlequin syndrome.

We opted for treatment with botulinum toxin (incobotulinumtoxin A) intradermally, conducted in two stages. In the first session, we marked the injection points all over the right hemiface with a spacing of about 1.5 cm between them (Figure 2). We used nine units of botulinum toxin and rediluted them in 27 volumetric units of 0.9% saline solution, totaling 36 volumetric units. At each point, 0.25 units of botulinum toxin were applied.

Two weeks later (Figure 1B), in a reassessment after the patient performed physical exercise for 20 minutes, we observed an improvement in the erythema and sweating on the right. We conducted a new marking in the areas with greater residual erythema. We applied four units of botulinum toxin rediluted in 12 volumetric units of 0.9% saline solution, totaling 16 volumetric units.

In a new reassessment, two weeks after the second application (Figures 1C and 3B), the patient showed a significant improvement in erythema and sweating on the right hemiface. There was no facial mime asymmetry after treatment (Figure 4).

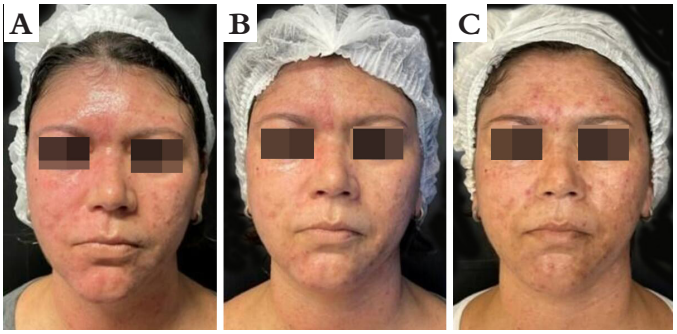

Figure 1: Comparative dermatological examination after performing physical activity for 20 minutes; **A -** Pre-treatment; **B -** Two weeks after the first application of botulinum toxin; **C -** Two weeks after the second application of botulinum toxin

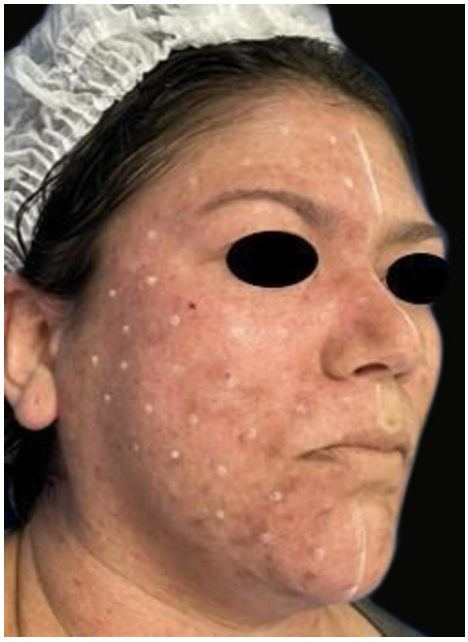

Figure 2: Marking of botulinum toxin application points in the first treatment session

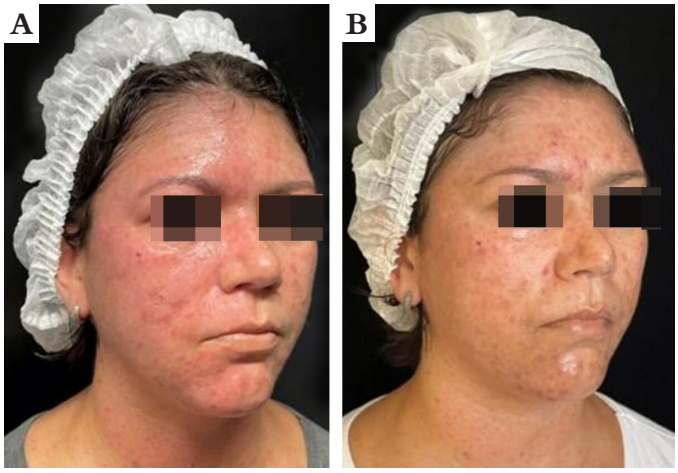

Figure 3: Comparative dermatological examination after performing physical activity for 20 minutes; **A -** Pre-treatment; **B -** Two weeks after the second application of botulinum toxin

DISCUSSION

Harlequin syndrome is idiopathic in most cases, but it can be congenital or secondary to structural or post-surgical iatrogenic lesions.[2] Therefore, it is essential to perform imaging tests to define the etiology.

The therapeutic approach considers the etiology. If structural damage is identified, targeted approaches are indicated. Otherwise, treatment aims at controlling the symptoms.

Ipsilateral sympathectomy is described as a surgical therapeutic possibility. However, the literature reports refractory cases. Stellate ganglion block is also reported as a treatment.[3] Oral medications, such as oxybutynin and propranolol, are therapeutic

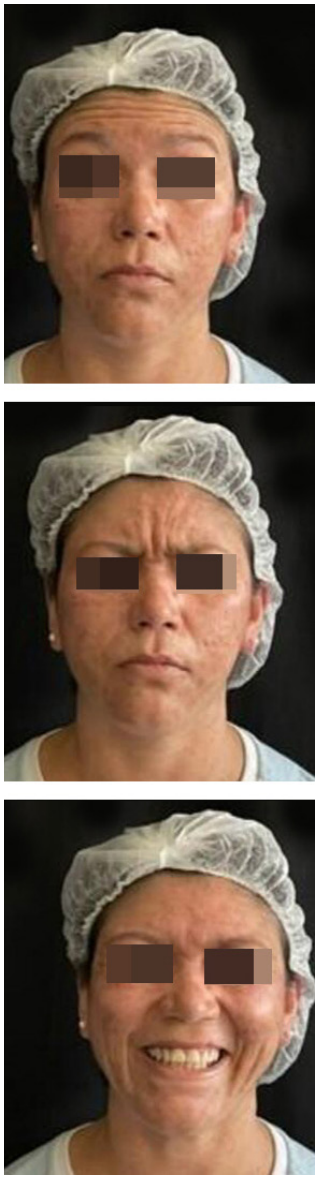

Figure 4: Absence of facial mime asymmetry after treatment

options,[4] but with little specificity to the affected areas and with more systemic adverse events.

Botulinum toxin application has been reported with good results. It prevents the release of acetylcholine, resulting in temporary chemo-denervation with loss or reduction of target organ activity. Therefore, it blocks the autonomic nerves of the vasodilator system.

We chose to perform treatment with botulinum toxin[5] as it is a minimally invasive, temporary, and targeted procedure, achieving promising results. ●

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