Casos Clínicos

Harlequin syndrome: a case report

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Resumen

El síndrome de Arlequín es una condición neurológica poco común, la mayoría de las veces benigna. Por lo general la mayor parte de los casos son de causa idiopática (aunque existen causas secundarias). Esta afección consiste en la aparición de eritema e hiperhidrosis facial unilateral desencadenado por el ejercicio, calor, emociones o estrés, la que se produce debido a una disfunción contralateral de la vía simpática vasodilatadora y sudomotora. Se presenta el caso de un paciente de sexo masculino de 49 años, que relata presentar un cuadro de 15 años de evolución caracterizado por la aparición de eritema y sudoración excesiva de la hemicara derecha, la cual termina abruptamente en la línea media de la cara, gatillado por el ejercicio y el calor en verano, mientras que la hemicara izquierda presenta anhidrosis sin eritema.

Palabras claves: síndrome de Arlequín; vía simpática; hiperhidrosis; eritema; anhidrosis.

Abstract

The Harlequin syndrome is a rare neurological condition, most of the time benign. In general, most cases are idiopathic (although there are secondary causes). This condition consists in the appearance of unilateral facial erythema and hyperhidrosis triggered by exercise, heat, emotions or stress, which occurs due to a contralateral dysfunction of the sympathetic vasodilator and sudomotor pathway. We present the case of a 49-year-old male patient, with no relevant morbid history, who reports a medical history of 15 years of evolution, characterized by the appearance of erythema and excessive sweating of the right side of his face, which ends abruptly in the middle line of the face, triggered by exercise and heat during the summer. While the left side presents anhidrosis without erythema.

Key words: Harlequin syndrome; sympathetic pathway; hyperhidrosis; erythema; anhidrosis.

Harlequin Syndrome (HS) is a rare autonomic neurological condition, considered a rare disease due to its low incidence, which is <1 / 1,000,000.[1] It is more common in women and is produced by a dysfunction in the upper thoracic sympathetic pathway, which causes redness and unilateral facial hyperhidrosis triggered by exercise, heat, emotion or stress.

HS was first described in 1988 by Lance et al.[2] The absence of unilateral facial redness and sweating is due to an ipsilateral dysfunction of the sympathetic vasodilator and sudomotor pathway, this alteration can occur at pre or post ganglionic level (Figure 1); It is postulated that compensatory sympathetic neuronal hyperactivity occurs on the healthy side, which accentuates the cutaneous signs of erythema and hemifacial hyperhidrosis that characterize this disorder.

Case Report

A 49-year-old male patient, resident in Santiago, came to our dermatology service on September 2020, for a mole checkup. Within his dermatological history, he presented a 15 – year clinical history of multiple episodes characterized by the appearance of erythema and excessive sweating in the right side of the face, which ends abruptly in the midline of the face, while the left side remains without erythema but presents anhidrosis. These episodes are triggered by exercise and heat during summer time. The patient reports that this does not occur during the winter months. When he does physical activity during winter time, only presents compensatory hyperhidrosis in the right side of his face. These episodes are self-limited from 40-45 minutes to 1 hour of duration, which spontaneously subside after exercise. He doesn't present other symptoms.

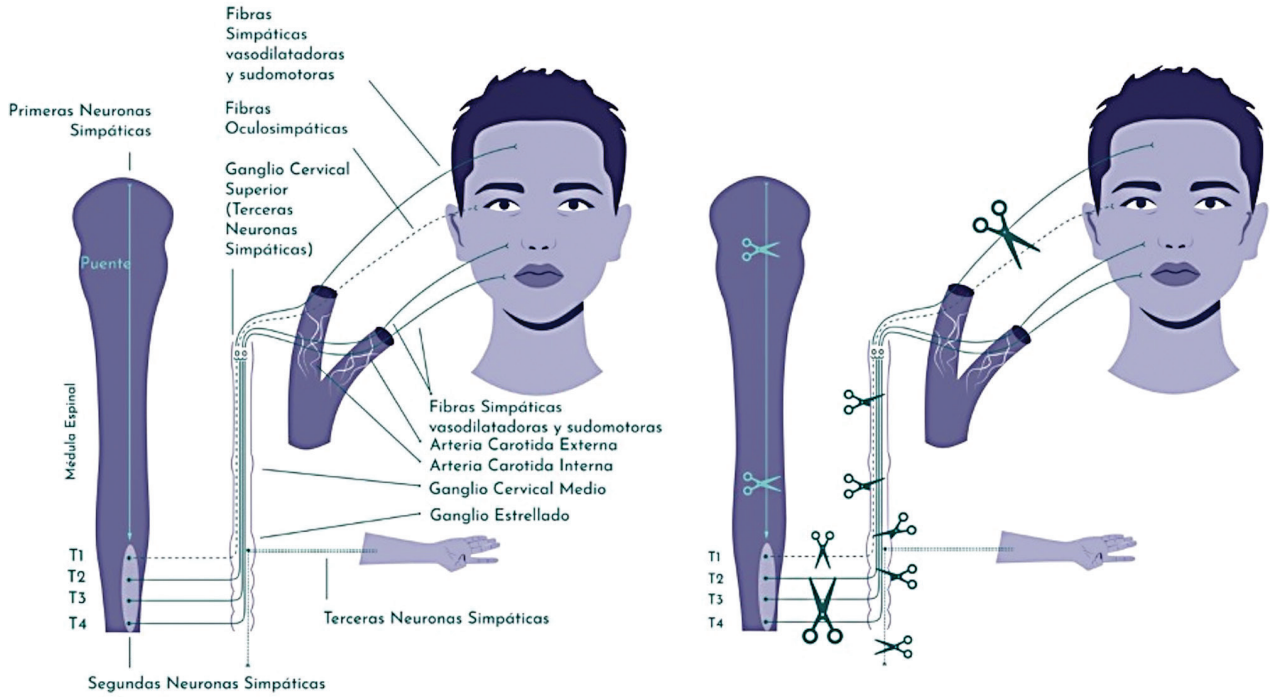

Figure 1
Harlequin Syndrome. Sympathetic outflow pathway: the first neurons originate from the hypothalamus and synapse in the spinal cord with the second (preganglionic) neurons. Most of the sudomotor and vasomotor fibers that innervate the face leave the spinal cord at T2-T4 level and travel to the superior cervical ganglion, where they synapse with the third (postganglionic) neurons that innervate the face. Scissors indicate possible sites of sympathetic lesions.[3]

Among his history, a non-staged renal failure and an allergy to NSAIDs stand out. He has no relevant family history. Within his medications he takes 100 mg allopurinol.

At the physical exam: patient of mesomorphic constitution, vital signs within normal ranges, normal cardiopulmonary examination, neurological examination: normal, no meningeal signs, no cerebellar signs. Skin: when resting there are no differences in color or asymmetries in the facial region. (Figure 2).

A clinical diagnosis of Harlequin Syndrome was made by a neurologist in April 2012.

In 2019, a total AP and lateral spine X-ray and an MRI of the dorsal spine were indicated, both had no pathological findings. In addition, he has an abdominal ultrasound scan and AP-L chest radiography, both from 2020, without pathological findings.

In view of the interest aroused by his syndrome, he was asked to send us a photo after jogging showing the erythema affecting the right side of the face. (Figure 3).

Figure 2
Harlequin Syndrome. Basal status

Figure 3
Harlequin Syndrome. Post exercise status

Discussion

Harlequin Syndrome (HS) is an infrequent neurological condition, considered a rare disease due to its low incidence, it is caused by a dysfunction of the sympathetic nervous system. It consists in the redness and unilateral facial hyperhidrosis triggered by exercise, heat, emotion or stress. In the case of our patient, the episodes are triggered by exercising during summer heat, generating erythema and hyperhidrosis of the right side of his face. When exercising during winter months, he has compensatory hyperhidrosis without erythema in the right side of his face.

Most of the cases of this rare condition are benign, mainly idiopathic, which is the case of our patient. In about 1 of 6 patients the disorder is caused by an underlying disease or structural injury (ie, secondary harlequin syndrome).[6] Some examples of secondary etiologies which can trigger this syndrome are: carotid dissection, toxic goiter, lung tumors, syringomyelia, multiple sclerosis and neuroblastoma.[5] There are also iatrogenic causes that have been increasing in recent years, as is the case with some procedures, for example: jugular catheterization and paravertebral anesthesia.[5]

HS is occasionally associated with some neurological disorders such as Horner syndrome, Adie syndrome and Ross syndrome, characterized by altered pupillary reflexes. When it is associated with any of these dysautonomic syndromes, it is called the Harlequin's sign.[6]

When making the diagnosis of a Harlequin syndrome, it is important to perform a detailed neurological physical examination where tendon reflexes and pupils are evaluated. In addition, different studies should be done to rule out any secondary underlying cause. Cervico-thoracic MRI, chest X-ray, brain MRI, and electrophysiological studies are recommended.[7,8]

In most cases, no treatment is required, unless there is a serious psychological impact on the patient that affects their self-esteem. In this case, a contralateral sympathectomy (interrupting facial redness and compensatory hyperhidrosis on the side of the face that does not have damage in the sympathetic pathway) should be considered, with the aim that both half-faces have a symmetrical forehead coloration against exercise or heat.[7] It's a surgery that's generally performed endoscopically, which can have complications, the most common is compensatory hyperhidrosis, which is generally well tolerated and decreases over the months. Other complications include: appearance of Horner syndrome, pneumothorax, hemothorax, infections, etc.[9]

The patient was talk through about the possibilities of further study and treatment, but he reports that his self-esteem or quality of life are not affected, so he does not want to undergo further tests or start any treatment.

Conclusion

Harlequin syndrome (HS) is a rare neurological disease, with a very low incidence, most of the time of benign idiopathic cause. It is produced by a dysfunction at any level of the upper thoracic sympathetic pathway.

When the diagnosis of this syndrome is made, imaging and electrophysiological studies should be carried out to rule out secondary causes that could be generating this disorder.

Because most of the time this syndrome is benign and idiopathic, it does not require any type of treatment. The interest in presenting this clinical case is given by its very low frequency and the fact that the patient corresponds to an idiopathic HS. Consideration should begiven to performing a contralateral sympathectomy in cases where the patient's self-esteem gets affected.

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