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Phantom eye syndrome. Case report

Síndrome de ojo fantasma. Reporte de caso

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What do we know about this issue?

- Phantom eye syndrome is a little reported pathology, although it is a frequent complication in the postoperative period of ophthalmic eye amputation surgeries.
- There are no specific treatment guidelines and the literary evidence on management is varied, as it is based on small studies and case reports.

What is this study's contribution?

- Evidence supporting the use of carbamazepine alongside other multimodal strategies that improved phantom pain, headaches and facial neuropathic pain in our patient.
- A brief summary of currently available literature, familiarizing us with the pathology's characterization, epidemiological data, risk factors and possible treatments.

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Abstract

A frequent complication after limb amputation is the appearance of phantom limb syndrome, a phenomenon that has also been studied in the amputation of different body parts. Cases reported in relation to ophthalmologic surgery are few and evidence is limited in terms of specific management, which makes this a very important study.

We report the case of a patient diagnosed with phantom eye syndrome in the post-operative period of an orbital exenteration surgery. A comprehensive approach was taken with multimodal symptom management, including intervention treatment. Significant improvement regarding the pain was achieved; however, non-painful phantom sensations persisted.

Upon reviewing the available literature on the subject, its pathophysiology is not fully elucidated. Its incidence is highly variable, as well as the symptoms' appearance. The presence of headache and eye pain prior to surgery seem to be risk factors. Climate and psychological stress are exacerbating factors of the symptomatology. No high-quality evidence was found in terms of management guidelines, with the use of antidepressants, anticonvulsants and opioids being the most recommended. Interventional management is an option according to the characteristics of the pain and associated symptoms.

Keywords

Pain; eye; phantom pain; phantom sensation; phantom limb; case reports; review.

Resumen

Una complicación frecuente luego de la amputación de alguna extremidad es la aparición del síndrome de miembro fantasma, fenómeno que también se ha descrito en la amputación de diferentes partes del cuerpo. Los casos reportados en relación con cirugía oftalmológica son pocos y la evidencia es limitada en cuanto a manejo específico, lo cual justifica la descripción de este caso de interés.

Reportamos el caso de una paciente a quien se diagnosticó síndrome de ojo fantasma en el periodo posoperatorio de una cirugía de exenteración orbitaria. Se realizó un enfoque integral con manejo multimodal de los síntomas, incluyendo tratamiento intervencionista. Se logró una notable mejoría del dolor; sin embargo, las sensaciones fantasma no dolorosas persistieron.

Al revisar la literatura disponible sobre el tema, la fisiopatología no está totalmente dilucidada. La incidencia es muy variable, así como el tiempo de aparición de los síntomas. La presencia de cefalea y dolor ocular previos a la cirugía parecen ser factores de riesgo. El clima y el estrés psicológico son factores exacerbantes de la sintomatología. No se encontró evidencia de alta calidad en cuanto a pautas de manejo, siendo lo más recomendado el uso de antidepressivos, anticonvulsivantes y opioides. El manejo intervencionista es una opción de acuerdo a las características del dolor y síntomas asociados.

Palabras clave

Dolor; ojo; dolor fantasma; sensación fantasma; miembro fantasma; informes de casos; revisión.

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INTRODUCTION

Phantom eye syndrome consists of any eye sensation a patient reports, even though the eye has been amputated. The syndrome includes: phantom vision, phantom pain and phantom sensations (1). It is different from both surgical wound pain and any other sensory disturbance in or around the surgical wound (2). Its prevalence varies according to the studies reported, being lower in relation to the phantom limb syndrome; however, given its complexity, we deem it very important to inform patients before surgery about this possible complication, as it can have a considerable impact on their quality of life (3). In this case report we present the approach and good response a patient had to the management described below.

CASE PRESENTATION

Patient information

A 69-year-old female, housewife, living in a rural area less than two hours from Bogotá, widowed, with five children and lives with her mother. Significant history of high blood pressure. She takes losartan, amlodipine and hydrochlorothiazide. She attended a consult at the pain and palliative care unit (PCU) of the Instituto Nacional de Cancerología in Bogotá for symptoms on her left eye, which had been amputated

by orbital exenteration. The symptoms were the sensations that the eye was still in place, eye movements and blinking (her eyelids were removed in the exenteration,) as well as burning pain on the eye surface and headache.

The symptom picture began two years ago with the appearance of a small pigmented lesion in the left eye's conjunctiva, associated with constant, mild occipital headache on the same side, for which she was not taking any painkillers. The ophthalmology service ordered three conjunctive biopsies in different surgical times. The last biopsy reported invasive melanoma. Orbital exenteration surgery was scheduled and was performed on October 29, 2018, without complications. From the immediate postoperative period, the patient presented phantom eye sensation, and two months later she started experiencing intermittent burning phantom pain of a 5/10 intensity in the Visual Analog Scale (VAS,) in addition to an increase in the intensity and frequency of the occipital headache on the left side already present before surgery and periorbital dysesthesia. This first consult took place five months after the surgery.

Clinical findings

Left orbit cavity exenterated through adequate re-epithelization process. Hypesthesia area at branches V1 and V2 of trigeminal nerve and absence of allodynia.

Calendar

Figure 1 presents the sequence of events in this patient's history.

Diagnosis

The first biopsy was taken on February 19, 2018 and the result was reported as undetermined, because it was not possible to define benign versus malignant lesion. The second biopsy was taken on March 26, 2018 and reported an atypical lesion, without possibility to determine malignant versus benign lesion. Up to that point, the differential diagnosis was of an atypical melanocytic lesion. Subsequently, the pathology of the third sample for biopsy was reported, which was taken on September 12, 2018: upper conjunctiva invasive melanoma (T2cN0M0.) The pathology of the product of the left eye exenteration of November 29, 2018 reported no residual invasion. Subsequently, in follow-up oncology controls, a cerebral nuclear magnetic resonance was prescribed, which was taken on December 19, 2018 and showed no alterations suggesting tumor recurrence. The computerized axial thorax and abdomen tomography taken on March 05, 2019 showed negative results for metastasis or relapse. CBC, renal function, liver function and electrolytes showed no alterations.

Interventions

After the first consult, the patient began taking 75 mg of pregabalin every 12 hours,

FIGURE 1. Timeline.



SOURCE: Authors.

which she took for a week but suspended due to intolerance. One month later, in the second consult, carbamazepine 200 mg was indicated at night, which was later modified in the third month to every 12 hours. At the same time, she took 1 g of acetaminophen every 8 hours and had ophthalmology, rehabilitation, plastic surgery and psychology controls.

Monitoring and results

The patient attended three more consults at the PCU in the sixth, seventh and ninth months of the postoperative period. In the second consult she reported persistent phantom sensations of frequent blinking of the exenterated eye, eye movement on the horizontal and vertical axes and phantom burning pain on the eyeball surface.

In the following consult she reported paresthesia and the appearance of lancinating, irruptive pain of an intensity of up to 7/10 (VAS) on the left periorbital area. Up to this point, she reported left occipital headache of an almost daily occurrence of up to 5/10 (VAS). The patient showed feelings of sadness and frustration, anxiety, constant crying and difficulty to remain asleep.

In the last consult, there was a good response to acetaminophen management of headaches, improvement in the intensity and frequency of the burning phantom pain, now of a 2/10 intensity (VAS,) since the start of carbamazepine. For periorbital neuropathic pain, interventional management with sphenopalatine block was performed. The procedure was carried out without complications and with very good response, with an 80% improvement of the pain.

PHANTOM EYE SYNDROME

In our patient's context, it is important to explain that orbital exenteration is a

radical procedure reserved for progressive neoplastic disorders or extensive facial trauma and consists of the extraction of the orbit's contents, including the periorbita. As it leads to disfigurement, it has a high psychological impact.

Pain sensation on an amputated limb was first recorded in the 16th century by French military surgeon Ambroise Paré. Such symptoms were described as phantom limb pain as early as in the 19th century (4-6). Today, very little is written about phantom eye syndrome, probably because the prevalence of orbital area cancer is very low (7). The etiology and pathophysiological mechanisms are not clearly defined. However, peripheral and central neural mechanisms have been described, along with overlapping psychological mechanisms (5).

Phantom pain is defined as all painful sensations referring to the missing limb or, as in this case, the missing organ (8,9). Phantom sensations are any sensation coming from the missing limb or organ, except for pain (6,8). Phantom manifestations have been described following tongue, teeth, breasts, penis and eye amputation (3,10). Phantom vision is a visual hallucination, a sensory experience with the convincing sense of reality of actual perception, but which occurs without external stimulation of the relevant sensory organ (1).

Epidemiology

Studies of phantom limb syndrome have identified that, after amputation, over 90% of patients may experience phantom phenomena (11); up to 80% may experience phantom pain and up to 70% may report symptoms even 25 years after amputation (3).

When talking specifically about oncological ocular pathology, the most frequent neoplasms that lead to amputation are choroidal melanomas in adults and retinoblastomas in children (1). Regarding these surgeries, the following have been

recorded as causes of postoperative pain: retrobulbar hematoma, prosthesis-related pain and pain of unknown origin (12).

In the reference study by Nicolodi et al. (7), a high incidence (71.6%) of phantom eye syndrome was observed in patients who underwent complete eye bulb enucleation. Symptoms appeared 7 days to 6 months after surgery, with peak incidence after 6 months. Andreotti et al. (2) found that up to 51% of patients who undergo eye amputation may suffer from phantom eye syndrome and up to 46% show at least one of its typical symptoms. In the study by Hope-Stone et al. (13), 60.3% of patients showed symptoms of phantom eye syndrome. For half of these patients, symptoms began within 6 weeks postoperatively, with phantom sensation being the most common and phantom vision or pain less so.

Phantom pain

The literature supports that the onset of pain is early. Several studies have shown that up to 75% of patients develop pain in the first few days after amputation. However, phantom pain can be delayed for months or years (13).

In the study by Sörös et al. (11), phantom eye pain prevalence was of 26%. Hope-Stone et al. (13) found that over one third of patients with phantom eye pain experienced symptoms daily. The episodes stopped spontaneously in 42.6% of the patients. Only a minority of patients were disturbed by their symptoms. However, those who experienced pain were more anxious and depressed.

These numbers suggest that phantom pain may be considerably less common after eye removal than after limb amputation (11).

In some patients phantom pain may gradually disappear over the course of a few months to a year, even without treatment, but other patients may have this pain for decades (4).

Phantom vision

Sörös et al. (11) found that the prevalence of phantom vision was of 31%. Visual hallucinations can be basic or complex. Onset typically occurs within a few days after surgery and the frequency usually decreases over time (2).

There are reports showing that these hallucinations may increase within a few weeks to 6 months after the eye is amputated (2).

The exact mechanisms of phantom vision are unknown. The involvement of plastic changes in the cerebral cortex inherent to vision, loss of physiological inhibition and hyperexcitation of the optic nerve stump has been suggested (11).

Phantom sensations

The prevalence of non-painful phantom sensations can be as high as 29% (11). Andreotti et al. (2) found that non-painful sensations are usually not observed immediately after surgery. Their highest incidence appears 2 to 12 months after surgery, and over time, the phantom sensation may disappear. Interestingly, the incidence of phantom sensations in the anophthalmic socket is low and manifests as itching around the eyes, a sensation of non-existent eyelids and a feeling of open eyelids. Most phantom sensations usually disappear after 2 to 3 years without treatment, except in cases where phantom pain develops (5).

Risk factors

The common factor in most studies is the relationship between headaches and eye pain as symptoms prior to surgery, with the appearance of phantom eye syndrome (2,7,11,13). These results seem to support the notion that patients with headaches have a dysfunctional sensory and painful signal transmission mechanism (7).

Cold, windy weather and psychological stress have been identified as the most common pain triggers (1,3).

Distress, anxiety and depression are also often correlated with an increase in phantom pain (10,13). Depression was diagnosed in 20 to 60% of amputees, a figure 3 to 5 times higher than among the general population (6). Anophthalmic patients have a lower quality of life compared to the general population, given the eyes' central role in communication and physical appearance. Eye removal can lead to devastating emotional issues, such as insecurity, inferiority complexes and fears of social marginalization (2).

Medical treatment

There are reported experiences with various medications for phantom limb pain, but there are no specific guidelines or optimal treatment for such cases. Opioids, N-methyl D-aspartate receptor antagonists, anticonvulsants, antidepressants, calcitonin and local anesthetics can be effective in improving symptoms such as pain, function, sleep pattern and quality of life. No treatment completely eliminates pain, and persistent long-term phantom pain can be resistant to any treatment (1,3,9).

Several studies found that morphine, gabapentin and ketamine provided short-term pain relief, but the findings were based primarily on small studies (9,14,15).

Intervention treatment

For neuropathic facial pain and related headache, the sphenopalatine ganglion is a promising treatment target through blocking, radiofrequency ablation and neurostimulation. Sphenopalatine ganglion blocking has some evidence supporting its use in some other circumstances (16). The strongest evidence refers to its use for cluster headache, migraine, trigeminal neuralgia and facial pain, reducing the need for pain medication after surgery (16). Blocking can provide

excellent pain relief with effect for up to 6 months, so it can be a potentially effective therapeutic tool (17).

DISCUSSION AND CONCLUSIONS

Phantom eye syndrome is a little reported but not infrequent pathology, with a high prevalence among orbital exenteration or enucleation postoperative complications (2,13). We do not have comparative data available in Colombian literature, hence the importance of this study.

In this clinical case, we highlight the identification of two of the syndrome's typical characteristics, such as phantom sensations and phantom pain (1,5,11); risk factors such as the presence of headache before surgery (7); and exacerbating factors such as emotional symptoms (10,13). Another important point was that, despite the absence of specific guidelines, there was a good response to the recommendations followed based on the available evidence (5,9,15), with an improvement of phantom pain and facial pain with neuropathic characteristics.

Some limitations were: the difficulty to differentiate a benign pathology from a malignant one in the first two biopsies' interpretation, and the permanence of non-painful phantom sensations despite the interventions performed. The symptoms' persistence is frequent and generates a substantial impact on the patients' quality of life (1,3).

This kind of report allows us to identify topics that can be studied further and should encourage the production of more structured studies that provide us with stronger data and conclusions. It is important that care divisions such as ophthalmology, pain, palliative care and rehabilitation do not let these types of complications go untreated, in order to generate specific guidelines for their management and approach.

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Contributions from the Authors

DSCD: Planning of the study, data collection, literature search, results interpretation, initial and final writing of the manuscript.

BMMA: Planning of the study, results interpretation, data analysis, final writing of the manuscript and final approval of the manuscript.

FALA: Case identification, original project conception, planning of the study, initial writing and final approval of the manuscript.

ETHICAL RESPONSIBILITIES

Protection of humans and animals

The authors declare that no experiments on humans or animals were conducted for this research.

Data confidentiality

The authors state that no patient data appears in this article.

Right to privacy and informed consent

The patient gave her express written consent authorizing the publication of his case description. This document is in the correspondence author's possession.

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Conflicts of interest

The authors state to have no conflict of interest regarding this paper.

REFERENCES

- Rasmussen MLR. The eye amputated - Consequences of eye amputation with emphasis on clinical aspects, phantom eye syndrome and quality of life. *Acta Ophthalmologica*. 2010;88:1–26. DOI: [10.1111/j.1755-3768.2010.02039.x](https://doi.org/10.1111/j.1755-3768.2010.02039.x).
- Andreotti AM, Goiato MC, Pellizzer EP, Pesqueira AA, Guiotti AM, Gennari-Filho H, et al. Phantom eye syndrome: A review of the literature. *Scientific World Journal*. 2014;2014: 686493. DOI: [10.1155/2014/686493](https://doi.org/10.1155/2014/686493).
- Rasmussen MLR, Prause JU, Toft PB. Phantom pain after eye amputation. *Acta Ophthalmol*. 2011;89 1:10–16. DOI: [10.1111/j.1755-3768.2010.02058.x](https://doi.org/10.1111/j.1755-3768.2010.02058.x).
- Kaur A, Guan Y. Phantom limb pain: A literature review. *Chinese J Traumatol*. 2018;21 6:366–368. DOI: [10.1016/j.cjtee.2018.04.006](https://doi.org/10.1016/j.cjtee.2018.04.006).
- Manchikanti L, Singh V. Managing phantom pain. *Pain Physician*. 2004;7:365-375.
- Pirowska A, Wloch T, Nowobilski R, Plaszewski M, Hocini A, Ménager D. Phantom phenomena and body scheme after limb amputation: a literature review. *Neurolog Neurochir Pol*. 2014; 48 1:52–59. DOI: [10.1016/j.pjnns.2013.03.002](https://doi.org/10.1016/j.pjnns.2013.03.002).
- Nicolodi M, Frezzotti R, Diadori A, Nuti A, Sicuteri F. Phantom eye: features and prevalence. The predisposing role of headache. *Cephalalgia*. 1997;17 4:501–504. DOI: [10.1046/j.1468-2982.1997.1704501.x](https://doi.org/10.1046/j.1468-2982.1997.1704501.x).
- Nikolajsen L, Jensen TS. Miembro fantasma. In: Wall y Melzack. *Tratado del Dolor*. Elsevier; 2007. p. 985–996.
- Fang J, Lian Y, Xie K, Cai S. Pharmacological interventions for phantom limb pain. *Chin Med J (Engl)*. 2013;126 3:542–549. DOI: [10.3760/cma.j.issn.0366-6999.20121599](https://doi.org/10.3760/cma.j.issn.0366-6999.20121599).
- Mccormick Z, Chang-Chien G, Marshall B, Huang M, Harden RN. Phantom limb pain: A systematic neuroanatomical-based review of pharmacologic treatment. *Pain Medicine*. 2014;15:292–305. DOI: [10.1111/pme.12283](https://doi.org/10.1111/pme.12283).
- Sörös P, Vo O, Husstedt I-W, Evers S, Gerding H. Phantom eye syndrome: Its prevalence, phenomenology, and putative mechanisms. *Neurology*. 2003;60 9:1542–1543. DOI: [10.1212/01.wnl.0000059547.68899.f5](https://doi.org/10.1212/01.wnl.0000059547.68899.f5).
- Shah-Desai SD, Tyers AG, Manners RM. Painful blind eye: efficacy of enucleation and evisceration in resolving ocular pain. *Br J Ophthalmol*. 2000;84 4:437–438. DOI: [10.1136/bjo.84.4.437](https://doi.org/10.1136/bjo.84.4.437).
- Hope-Stone L, Brown SL, Heimann H, Damato B, Salmon P. Phantom Eye Syndrome: Patient Experiences after Enucleation for Uveal Melanoma. *Ophthalmology*. 2015;122 8:1585–1590. DOI: [10.1016/j.ophtha.2015.04.005](https://doi.org/10.1016/j.ophtha.2015.04.005).
- Alviar MJM, Hale T, Dungca M. Pharmacologic interventions for treating phantom limb pain. *Cochrane database Syst Rev*. 2016;10: CD006380. DOI: [10.1002/14651858.CD006380.pub3](https://doi.org/10.1002/14651858.CD006380.pub3).
- Alviar MJM, Hale T, Dungca M. Pharmacologic interventions for treating phantom limb pain. *Cochrane database Syst Rev*. 2011;12:CD006380. DOI: [10.1002/14651858.CD006380.pub2](https://doi.org/10.1002/14651858.CD006380.pub2).
- Ho KWD, Przkora R, Kumar S. Sphenopalatine ganglion: block, radiofrequency ablation and neurostimulation - a systematic review. *J Headache Pain*. 2017;18:118. DOI: [10.1186/s10194-017-0826-y](https://doi.org/10.1186/s10194-017-0826-y).
- Lee SH, Kim Y, Lim TY. Efficacy of sphenopalatine ganglion block in nasal mucosal headache presenting as facial pain. *Cranio*. 2018;8:1–3. DOI: [10.1080/08869634.2018.1475859](https://doi.org/10.1080/08869634.2018.1475859).