

# Squamous Cell Carcinoma of the Pinna in a Patient with Jorge Lobo Disease: Case Report

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*Carcinoma Espinocelular de Pavilhão Auricular em Paciente com Doença de Jorge Lobo: Relato de Caso*

Carcinoma de Células Escamosas del Pabellón Auricular en Paciente con Enfermedad de Jorge Lobo: Informe de Caso

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## ABSTRACT

**Introduction:** Lobomycosis (also called Jorge Lobo's Disease) is a chronic granulomatous disease caused by the fungus *Lacazia loboi*, affecting mainly rural workers, generally after local trauma to exposed skin areas, more common in men, aged 20 to 40, characterized by slow progression over the years, of varied skin lesions, notably nodules and coalescent plaques with a keloid appearance. It is known that chronic lesions such as these can become malignant, resulting mainly in squamous cell carcinoma. After analyzing skin cancers, it is suggested that lobomycosis also correlates with carcinogenesis, although it is little described in the literature and a complete elucidation has not yet been achieved. **Case report:** Male patient, 63 years old, born in Pará, rural worker, suffering from lobomycosis with a previous history of basal cell carcinoma with squamous differentiation in the right earlobe and pre-auricular region, having undergone resection of the lesion with margins in 2018, but evolving after nearly three years with the appearance, on the same sites, of a new tumor lesion, this time with greater aggressiveness and pathology compatible with squamous cell carcinoma. **Conclusion:** Considering the rarity of lobomycosis in non-Amazonian areas, the case in question is essential to demonstrate that this condition can be associated with skin neoplasms, contributing as a risk factor due to the formation of scars and chronic ulcers, in addition to warning on the need for early diagnosis and treatment to avoid carcinomatous degeneration.

**Key words:** Lobomycosis; Carcinoma, Squamous Cell; Skin Neoplasms.

## RESUMO

**Introdução:** A lobomicose (também chamada de doença de Jorge Lobo) é uma doença granulomatosa crônica, causada pelo fungo *Lacazia loboi*, acomete principalmente trabalhadores rurais, geralmente após traumas locais em áreas cutâneas expostas. É mais comum em homens, de 20 a 40 anos, caracterizando-se por progressão lenta, no decorrer dos anos, de lesões cutâneas variadas, notadamente nódulos e placas coalescentes de aspecto quelóideano. Sabe-se que lesões crônicas como essas podem malignizar, resultando principalmente no carcinoma espinocelular. Ao analisar os cânceres de pele, sugere-se que a lobomicose também tem correlação com a carcinogênese, embora seja pouco descrita na literatura e uma elucidação completa ainda não tenha sido traçada. **Relato do caso:** Homem, 63 anos, natural do Pará, trabalhador rural, portador de lobomicose e com história prévia de carcinoma basocelular com diferenciação escamosa em lóbulo da orelha e região pré-auricular direitos. Ele foi submetido à ressecção da lesão com margens em 2018, porém evoluiu após cerca de três anos com o surgimento, nas mesmas topografias, de nova lesão tumoral, desta vez com maior agressividade e com anatomopatológico compatível com carcinoma espinocelular. **Conclusão:** Tendo em vista a raridade da lobomicose em áreas não amazônicas, o caso em questão é fundamental para evidenciar que essa afecção pode associar-se com neoplasias de pele, contribuindo como fator de risco em razão da formação de cicatrizes e úlceras crônicas, além de alertar para a necessidade de diagnóstico e tratamento precoces, a fim de evitar a degeneração carcinomatosa.

**Palavras-chave:** Lobomicose; Carcinoma de Células Escamosas; Neoplasias Cutâneas.

## RESUMEN

**Introducción:** La lobomicosis (también llamada enfermedad de Jorge Lobo) es una enfermedad granulomatosa crónica, causada por el hongo *Lacazia loboi*, que afecta principalmente a hombres trabajadores rurales, generalmente después de traumatismos locales en áreas expuestas, de 20 a 40 años, caracterizada por progresión lenta durante años de diversas lesiones cutáneas, en particular nódulos y placas coalescentes de aspecto quelóide. Se sabe que lesiones crónicas como estas pueden volverse malignas, resultando principalmente en carcinoma de células escamosas. Al analizar los cánceres de piel, se sugiere que la lobomicosis también tiene correlación con la carcinogénesis, aunque está poco descrita en la literatura y aún no se ha dilucidado por completo. **Informe del caso:** Hombre, 63 años, nacido en Pará, trabajador rural, con lobomicosis y antecedentes de carcinoma basocelular con diferenciación escamosa en lóbulo de oreja y región preauricular derechas, sometido a resección de la lesión con márgenes en 2018, pero evolucionando después de tres años con la aparición, en las mismas topografías, de una nueva lesión tumoral, esta vez con mayor agresividad y compatible en el examen anatomopatológico con carcinoma de células escamosas. **Conclusión:** Considerando la rareza de la lobomicosis en zonas no amazónicas, el caso es fundamental para demostrar que esta condición puede estar asociada a neoplasias de la piel, contribuyendo como factor de riesgo por la formación de cicatrizes y úlceras crónicas, además de alertar a la necesidad de un diagnóstico y tratamiento precoces para evitar la degeneración carcinomatosa.

**Palabras clave:** Lobomicosis; Carcinoma de Células Escamosas; Neoplasias Cutáneas.

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## INTRODUCTION

Jorge Lobo's disease, also known as lacaziosis or lobomycosis, originally described in Recife, Pernambuco, by the dermatologist Jorge Lobo in 1931, is a chronic granulomatous disease, caused by the fungus *Lacazia loboi*<sup>1</sup>, that targets mainly rural workers or people who work in direct contact with the soil or recent travel to forest regions, such as the Amazon<sup>2</sup>. The incubation period is believed to be from one to two years after the fungus invades traumatic skin injuries. The infection manifests through different skin lesions, such as keloid nodules, ulcers, or warty lesions, and can slowly progress to several parts of the body through the years<sup>2</sup>.

In the same theme, squamous cell carcinoma is characterized by the abnormal and uncontrolled growth of squamous cell in the skin, with a complex and multifactorial physiopathology. The tumor usually presents as a crusty and coarse skin lesion, preferably developing in sun-exposed areas, such as the face, neck, ears and upper limbs<sup>3</sup>.

Records of skin tumors in patients with Jorge Lobo's disease are very rare in the literature<sup>1</sup>. This article presents a case of voluminous squamous cell carcinoma that spans from the right pinna until the ipsilateral pre-auricular region, in a patient with Jorge Lobo's disease and a history of basal cell carcinoma in the right earlobe and pre-auricular region, to illustrate and discourse about a case where malignancy might have occurred through a lobomycosis granulomatous lesion or even the association of this rare affection with the process of carcinogenesis, considering the related risk factors, emphasizing the importance of early diagnosis and treatment of lobomycosis, as well as its follow-up.

The present study has been approved by the *Hospital Geral de Fortaleza* (HGF) Research Ethics Committee, approval report number 6.646.738 (CAAE (submission for ethical review): 76900523.0.0000.5040), in compliance with Resolution 466/2012<sup>4</sup> of the National Health Council. The clinical case was correlated to the specialized literature on the theme and a bibliographical review on the subject was done using the following databases: SciELO, PubMed and Capes journal portal.

## CASE REPORT

Man, brown skin, 63-years old, from the town of Cratêus, state of Ceará, born in the state of Pará, rural worker, high blood pressure, chronic non-dialysis kidney disease and longtime carrier of Jorge Lobo's disease, with

a history of basal cell carcinoma resection with squamous differentiation in the right earlobe and pre-auricular region, with graft rotation, in 2018, in the *Hospital Haroldo Juaçaba*. Started follow-up in the HGF in 2022, due to new tumoral lesion in the same topography, extended to the pinna, external auditory canal and inferior crura of the antihelix (Figure 1A).

The patient had recent histopathological of the referred lesion showing infiltrating solid carcinoma with reticulated areas and signs of squamous differentiation with keratin pearls, with the indication of surgical treatment. Moreover, he showed ulcerated lesions in the left lower limb (Figures 1B and 1C), as well as multiple keloid lesions disseminated through the whole body, mainly the lower limbs (Figure 1D). The patient had a computerized tomography (CT) of the neck, with contrast, one month before the surgery (Figures 2A and 2B), which evidenced an expansive lesion with no well-defined cleavage plan.

During the operation, a frozen section biopsy of the lesion in the right pre-auricular region and

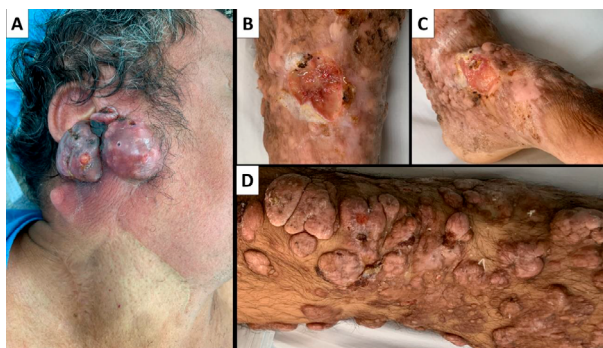


Figure 1. A. Tumoral lesion in the right pinna spread to the pre-auricular region on the day of operation. B. Highlight to the ulcerated lesion in the anterior face of the distal portion of the left leg. C. Highlight to the ulcerated lesion on the left ankle. D. Keloid-like plaques spread throughout the right lower limb.

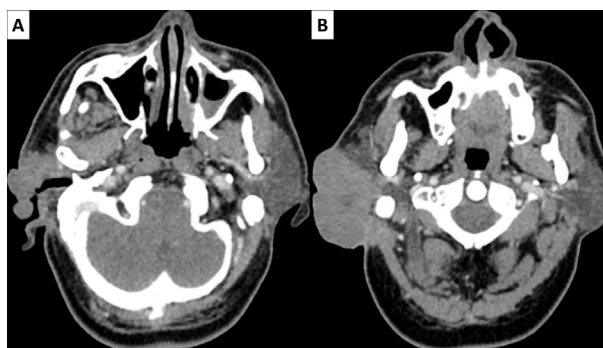


Figure 2. A. Heterogeneous infiltrative expansive lesion with a neoplastic appearance, affecting the superficial and deep adipose planes of the periauricular region. B. Extension of the neoplastic lesion up to the superficial lobe of the right parotid gland and near the insertion of the sternocleidomastoid muscle, without a well-defined cleavage plane.

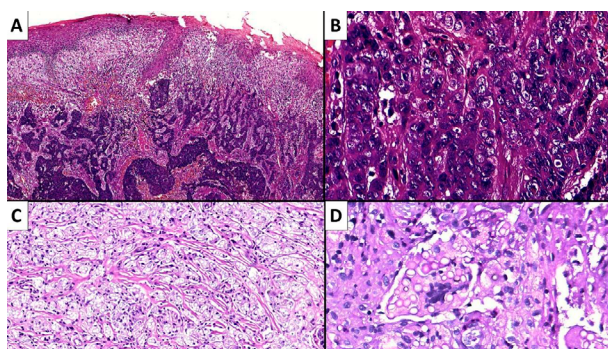
pinna was performed, which confirmed the squamous cell carcinoma, therefore prompting a full resection of the whole lesion with margins, in addition to supraomohyoid neck dissection and rotation of the right pectoralis major muscle flap, with placement of cervical and thoracic drains, as well as biopsy of ulcerated lesions in the lower limbs.

As the patient's respiratory tract was considered very difficult by the anesthesia team and there were risks of evolving to acute respiratory failure, due to the broad surgical manipulation of the right cervical region, the anesthetist opted to take the patient intubated to the postoperative ICU. Due to the difficulty in weaning from ventilation and the risk of complications from possible extubation failure, the head and neck surgery team performed an early tracheotomy, two days after the surgery.

Analysis of the lesion that extended from the right pre-auricular region to the ipsilateral pinna, measuring 7.0 x 4.2 x 4.3 cm, was compatible with histological grade II, moderately differentiated (Figures 3A and 3B), with invasion into the dermis, free circumferential margins and compromised deep margin, with macroscopic involvement of soft tissues, muscle, parotid gland and auricular cartilage.

Microscopy of the ulcerated lesions on the left lower limb showed intense macrophagic infiltration parasitized by oval microorganisms, usually of homogeneous size (approximately 10 micrometers), all intracellular and presenting catenulate arrangements, suggestive of lobomycosis, with areas of associated fibrosis, without signs of neoplasm.

The patient remained hospitalized in the ICU for about a month, having evolved with difficulty in



**Figure 3.** A. Histological section of the tumor lesion in the right pinna and pre-auricular region, compatible with histological grade II squamous cell carcinoma, moderately differentiated, with invasion into the dermis (hematoxylin-eosin 20x). B. Histological section evidenced in A with greater increase (hematoxylin-eosin, 40x). C. Histological section of ulcerated lesions on the patient's left lower limb, showing catenulate arrangements in a pattern known as a "pearl necklace", characteristic of lobomycosis (PAS, 20x). D. Higher magnification histological section of lesion in C, showing multinucleated cell with large number of fungi inside (PAS, 40x).

weaning from ventilation and pneumonia associated to mechanic ventilation, with *Pseudomonas aeruginosa* isolated in tracheal aspirate. The patient recovered from the infectious condition, however, needed hemodialysis after using broad antimicrobial regimen. The treatment for Jorge Lobo's disease started during the mentioned hospitalization, using clofazimine and itraconazole, according to guidance by the infectious disease specialist.

In parallel, the patient showed good postoperative recovery, with flap viability and joint monitoring with the stomatology team regarding open areas due to the extensive surgical failure after removal of an extensive tumor lesion, in order to make the pectoralis major muscle flap viable. Finally, the patient showed clinical improvement, the tracheotomy was removed, and he was discharged from the hospital after approximately two months of hospitalization, for outpatient follow-up.

In subsequent follow-ups, after complete healing of the surgical wound and regression of the cervical edema, the patient was referred to radiotherapy in a referral hospital for oncological diseases. Audiometry, performed before radiotherapy, showed bilateral moderate mixed hearing loss. The patient is still following-up with the otorhinolaryngology and head and neck surgery wards of HGF.

## DISCUSSION

Jorge Lobo's disease predominates in men aged 20 to 40 years-old and is characterized by the appearance of polymorphic lesions with slow growth. This disease encompasses nodules that resemble keloids, ulcerated or with warty plaques appearance, notably in trauma locations and exposed body parts, such as ears, face, arms, legs, chest, with no reported case in mucous membranes. The lesions of this disease disseminate through proximity as well as through self-inoculation and through the lymphatic pathway<sup>1</sup>.

The physiopathology of lobomycosis is still little understood; however, it can be associated to a chronic inflammatory response of the immune system to the fungus *Lacazia loboi*. The fungal infection in question leads to the formation of granulomas, that originate from the attempt to encapsulate the microorganism in order to prevent its spread. The fungi are found mainly inside macrophage vacuoles, reproducing mainly by budding, forming linear chains with a birefringent cell wall that contains melanin<sup>2</sup>.

Differential diagnoses for Jorge Lobo's disease include leprosy, leishmaniasis, sporotrichosis, paracoccidioidomycosis, blastomycosis, histoplasmosis,



Kaposi's sarcoma, cutaneous tuberculosis, benign and malignant tumors, cutaneous metastases, among others<sup>1</sup>. The lobomycosis diagnosis is performed through anamnesis and macroscopic analysis of lesions, in addition to direct microscopy, culture and histopathological evaluation<sup>2</sup>.

Malignancy of skin lesions was initially reported since the 19th century, in burn sequelae, by Jean-Nicholas Marjolin. In this context, malignancy of ulcerated lesions from chronic ulcers and scar lesions of several diverse etiologies, including those of an infectious nature, such as lacaziosis, are known as "Marjolin's ulcers"<sup>5</sup>.

Old scars and chronic ulcers of any kind can lead to the development of squamous cell carcinoma, having been described by many authors in patients with chronic granulomatous diseases, though the specific association with Jorge Lobo's disease is not so evident in the literature<sup>6</sup>. Squamous cell carcinoma is the most common skin cancer in those cases and results from the malign proliferation of keratinocytes<sup>7</sup>.

Chronic exposure to ultraviolet radiation (UV), coming from solar beams, is one of the main risk factors for the development of squamous cell carcinoma, inducing mutations in the DNA of epidermal cells, resulting in the promotion of carcinogenesis. Moreover, it is also possible to correlate this neoplasm to the following risk factors: smoking, human papillomavirus (HPV) infection, light skin, old age, exposure to carcinogenic chemical substances (like arsenic) and genetic predisposition<sup>8</sup>.

The chosen treatment for Jorge Lobo's disease ranges from systemic antifungal, electrocoagulation, cryotherapy or even surgical excision with wide margins, depending on the clinical manifestation, but recurrence is frequent, despite any therapy, in addition to notable aesthetic and functional impairment<sup>6</sup>.

## CONCLUSION

Since Jorge Lobo's disease is not well known, especially in non-Amazonian regions, and most cases happen in the country, the present article has an important role of showcasing this affection in a patient that presented squamous cell carcinoma in the same topography where, three years prior, a basal cell carcinoma was diagnosed with squamous differentiation. It is essential to understand the chronic nature of this fungal disease, which leads to the formation of granulomas in the most diverse parts of the body, which can evolve into ulcers and culminate in a malignant process, increasing morbidity and stigma in a condition that already has very marked dermatological manifestations.

The case report inquires if the initial lesion in the right pinna was due to Jorge Lobo's disease and whether this evolved into an ulcer that later resulted in skin carcinomas, or whether lobomycosis acted only as a risk factor, fueling local carcinogenesis, due to the persistent inflammatory stimulus of this fungal disease, which in the patient portrayed did not receive adequate monitoring until the development of basal cell carcinoma.

Thus, early diagnosis and treatment, as well as following up with these patients, is very important to prevent future neoplasms and its consequences.

## CONTRIBUTIONS

All the authors have substantially contributed to the study design, acquisition, analysis and interpretation of the data, wording, and critical review. They approved the final version for publication.

## DECLARATION OF CONFLICT OF INTERESTS

There is no conflict of interest to declare.

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None.

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