Extraocular sebaceous carcinoma of the scalp: case report and therapeutic challenges

Carcinoma sebáceo extraocular de cuero cabelludo: reporte de caso y desafíos terapéuticos

Larissa Mirna Belén Campuzano Ortíz*, Evelio Legal Balmaceda**, Lisa Carolina Arguello***, Marta Elizabeth Osorio****

Instituto de Previsión Social, Hospital de Especialidades Quirúrgicas – Ingavi. Servicio de Cirugía General. Fernando de la Mora, Paraguay

ABSTRACT
Sebaceous gland carcinoma (SGC) is a rare and aggressive malignant cutaneous neoplasm. 75-80% originate in the periocular region. Extraocular location, particularly on the scalp, is extremely rare. We present the case of a patient with scalp SGC, initially diagnosed as a sebaceous cyst. Treatment was based on surgical resection and adjuvant radiotherapy. Diagnostic and therapeutic challenges are discussed. The prognosis is guarded due to the high rate of recurrence and metastasis.[1]

Keywords: Sebaceous carcinoma, Extraocular sebaceous adenocarcinoma, Muir-Torre syndrome, scalp.

RESUMEN
El carcinoma de glándula sebácea (CGS) es una neoplasia cutánea maligna agresiva y poco frecuente. El 75-80 % se origina en la región periocular. La localización extraocular, particularmente en cuero cabelludo, es extremadamente rara. Se presenta el caso de una paciente con CGS de cuero cabelludo, inicialmente diagnosticado como quiste sebáceo. El tratamiento se basó en resección quirúrgica y radioterapia adyuvante. Se discuten los desafíos diagnósticos y terapéuticos. El pronóstico es reservado debido a la alta tasa de recurrencia y metástasis. [1]

Palabras claves: Carcinoma sebáceo, Adenocarcinoma sebáceo extraocular, síndrome de Muir-Torre, cuero cabelludo.

INTRODUCTION

Sebaceous gland carcinoma (SGC) is a rare, malignant, aggressive cutaneous neoplasia originating from the sebaceous glands.[1] In most cases (75-80%) it presents itself on the periocular region, mainly on the eyelids, deriving from meibomian or Zeis glands. Extraocular localizations are extremely rare, with head and shoulders being the more common ones within these unusual presentations.[3]

SGC's etiology isn't completely defined. Mutations in the p53-tumors-suppressing gene have been identified in some cases. Other risk factors include immunosuppression (organ transplant, HIV infection), previous radiotherapy and Muir-Torre syndrome (MTS). The latter is a dominant autosomal genetic syndrome characterized by the presence of sebaceous tumors (adenomas, epitheliomas or carcinomas) and at least one visceral neoplasia, more commonly known as colorectal cancer. [2] Among individuals with Muir-Torre syndrome, greater exposition to environmental ultraviolet radiation is associated with a larger risk of sebaceous carcinoma, as well being an independent risk factor for this type of cancer.[3]

SGC mainly affects elder adults (60-80 years old), predominantly women and mostly in Asian populations. Clinically, ocular presentation tends to be a yellowish node on the upper eyelid. Extraocular injuries, on the other hand, manifest themselves as unspecific, fast-growth, pink or red-yellowish nodes.[2] Definitive diagnosis requires biopsy and a histopathological study.

Due to the rarity of the SGC's extraocular presentation on the scalp, and the diagnostic and therapeutic challenges it entails, we present the following clinical case.

The treatment of choice for the SGC is complete surgical excision. Mohs micrographic surgery is an option that allows for precise control of the margins, especially useful on the periocular region. Sentinel node biopsy can be considered in periocular tumors but isn't routinely recommended for extraocular localizations due to the low node metastasis rate. [4]

Adjuvant radiotherapy, with 50-60 Gy doses, is indicated in perineural invasion, positive nodes cases or positive surgical margins. In unresectable or advanced with microsatellite instability or high mutational charge tumors, the pembrolizumab

* General Surgery Specialist. Nuestra Señora de la Asunción's Catholic University (Universidad Católica Nuestra Señora de la Asunción). Social Prevision Institute.
** General Surgery Specialist. National University of Asunción (Universidad Nacional de Asunción). Otolaryngology Subspecialist. Masters in oncologic surgery, San Pau. Barcelona, Spain.
*** Oncologic Surgery Specialist. National Cancer Institute (Instituto Nacional del Cáncer). Oncologic Otolaryngology.
**** Oncologic Surgery Specialist. National Cancer Institute (Instituto Nacional del Cáncer). Heliópolis Hospitalary Complex (Complejo Hospitalar Heliópolis), San Paulo, Brazil.
Corresponding Author: Dra. Larissa Mirna Belén Campuzano Ortíz, Address: Republica de Colombia casi Chile. Mariano Roque Alonso, Paraguay.
Email: larissacampuzano1@gmail.com
Received: 08/10/2024 - Revised: 20/10/2024 - Accepted: 04/11/2024
Revisor: Nelson Martínez ORCID - Affiliation: Faculty of Medical Sciences UNA
Editor: Eduardo González Miltos National University of Asunción (Universidad Nacional de Asunción) San Lorenzo, Paraguay

(a PD-1 inhibitor) has proven to be efficient.[5-6]

CASE PRESENTATION

40-year-old female patient, no relevant pathological history. Denies toxic habits.

Reported due to scalp injury, left parietal region, with a 10-month evolution. Initially, the injury was diagnosed as a sebaceous cyst in another center, having a local excision and antibiotic therapy performed. Five months after, the patient noticed a recurrence of the injury on the same site, with progressive growth, ulceration, bleeding and pus secretion.

She reported to the urgency service in a well general state, hemodynamically stable and afebrile. The physical exam yielded an erythematous, ulcerated tumor of irregular edges, with pus secretion, consistent, not painful to touch, of approximately 6 cm (2.36 inches) of major diameter, localized on the left parietal region of the scalp *(Figure 1 y 2)*. No cervical adenopathies found upon touch.

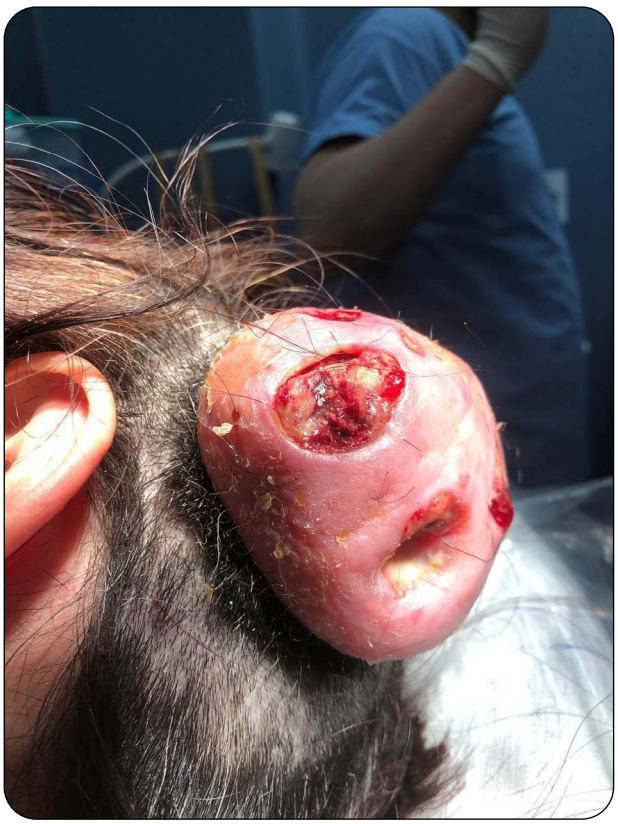

Figure 1. Preoperatory injury aspect.

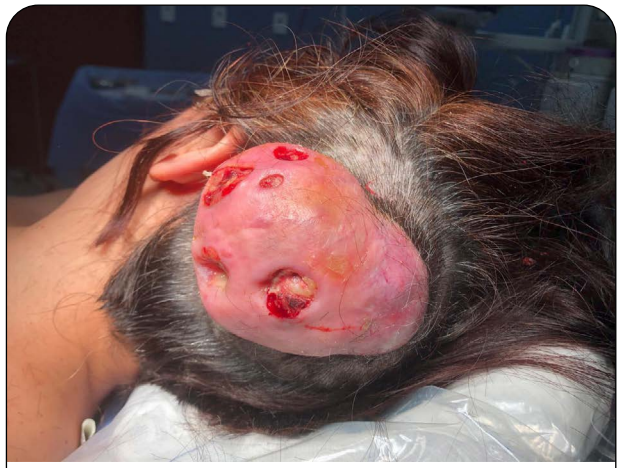

Figure 2. Presurgical injury.

Laboratory studies yielded no sign of systematic infection. 4800/mm3 white blood cells, 55% neutrophils, 12.2g/dL hemoglobin, 37.6% hematocrit, 275,000/mm3 platelets, 1.2 PCR. Computerized tomography (CT scan) of the cranium revealed an absence of lytic or blastic injuries which would suggest bone invasion. An exophytic lesion is identified in the scalp, of heterogenous density, ill-defined edges on the left parietal region. The first impression is to present a wide implantation on the galea. *(see Figure 3 y 4)*

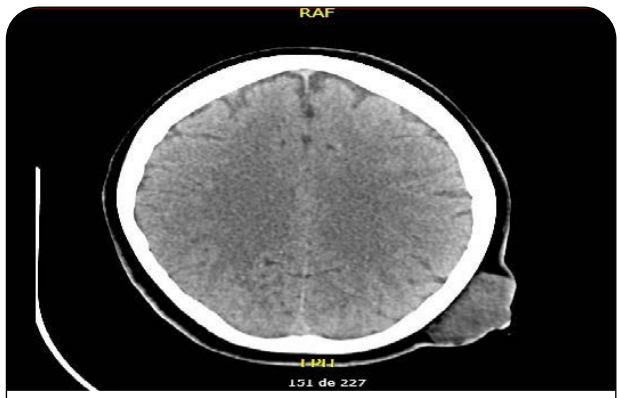

Figure 3. Preoperatory tomography.

Figure 4. Postoperative injury.

An excisional biopsy was performed under general anesthesia. A rhomboid rotational flap was used to reconstruct the defect, due to the size of the tumor and the need to preserve the scalp's vascularization. The primary cutaneous defect was designed in a rhomb shape with four equal sides and two 120° angles and two 60° ones. Usually, four potential flaps can be extracted from a rhomboid defect. The incision was performed, including margins of 2 cm (0.78 inches), and subgaleal dissection, encompassing pre-established margins. A 6 cm (2.36 inches) tumor was identified, friable, ulcerated, with slight adherence to the galea, which has completely dried. Hemostasia was performed, washing with a saline solution and oxygenated water, and a Penrose-type laminar drainage was placed. Closure was performed with polyglactin 3-0 suture for the subcutaneous cell tissue by simple stitching and nylon 3-0 for the skin by simple stitching.

During the immediate postoperative, partial undoing of the injury was observed. Daily healing was performed with a saline solution, achieving a favorable evolution. The patient was discharged after three days of in-patient stay.

The anatomopathological study yielded a unifocal sebaceous carcinoma, G2-G3 histological grade (moderately to poor differentiated), without lymph-vascular nor perineural invasion, although with surgical margin compromise.

The case was discussed in a multidisciplinary committee and adjuvant radiotherapy was decided upon. A cervical echography pre-radiotherapy discarded metastatic adenopathies. After the administration of two radiotherapy sessions, through a three-month follow up, the patient was evaluated in the service of external consultation. She remained asymptomatic, with adequate scarring of the injury *(see Figures 4 y 5)*. Tomographic control of the cranium and neck was performed, yielding an absence of the previously documented injuries. Comparative analysis shows complete resolution of the anomalies in presurgical studies. *(Figure 5)*.

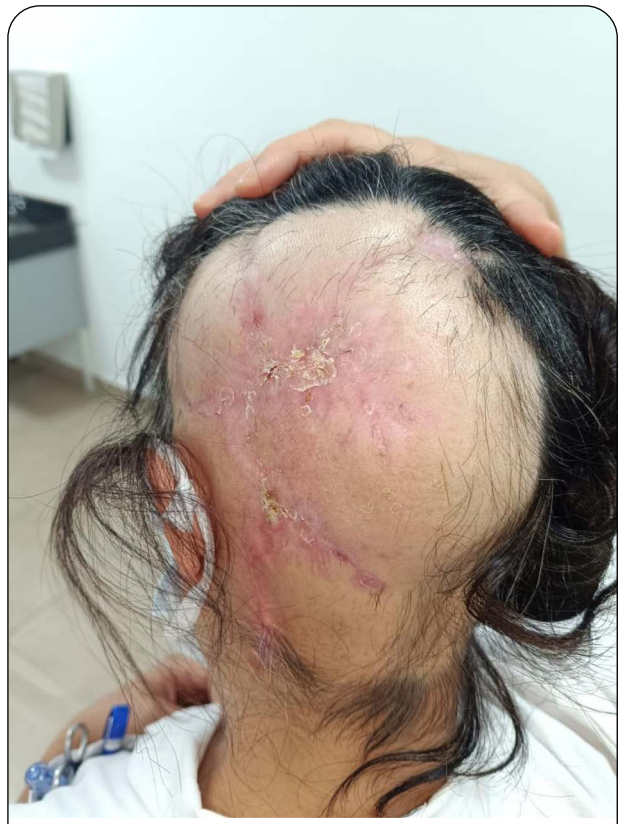

Figure 5. Scarring of the surgical area.

The findings match a satisfactory postoperative evolution, with no evidence of recurring injury or acute complications.

DISCUSSION

This case presents an unusual manifestation of an extraocular SGC in the scalp. Extraocular localization represents only 20-25% of all SGC's, as it's mentioned by MA Martin and collaborators. [7] Of these, a small fraction is presented in the scalp. The wrongful initial diagnosis as a sebaceous cyst, followed by a recurrency, highlights the importance of considering SGC during the differential diagnosis of scalp cutaneous injuries, including those in young patients and without evident risk factors, matching other published works. [8]

A multidisciplinary approach, with surgical, medical oncology and radiotherapy participation is crucial to the optimal handling of these tumors as pointed out by Jimenez [9]. Complete surgical resection, with wide margins, is the pillar of the treatment. In this case, the selection of the rhomboid-type rotative flap allowed for adequate reconstruction of the defect, preserving the vascularization and minimizing tension.

Adjuvant radiotherapy was indicated due to the compromise in a surgical frame, despite the absence of lymph-vascular or perineural invasion. This decision is based on the high local recurrency rate of SGC, even after an apparent complete resection. NCCN (National Comprehensive Cancer Network) guidelines recommend adjuvant radiotherapy in positive margin cases, perineural invasion or node disease.[10]

The limitations of this case include short-term follow up and

the lack of gene studies.

CONCLUSION

This case illustrates the unusual presentation of an extraocular sebaceous carcinoma in the scalp and the importance of a multidisciplinary approach for its handling. Despite this neoplasia's rareness, it's fundamental to consider it during the differential diagnosis of cutaneous injuries in this area, especially in patients with risk factors. Complete surgical resection with wide margins, followed by adjuvant radiotherapy in selected cases, offers the best chance of locoregional control. A long-term follow up is necessary to detect recurrence or metastasis.

CONFLICT OF INTERESTS

Authors declare no conflict of interests. Compliance with ethical norms.

REPORTED CONSENT

Reported consent was obtained from the patient for the case presentation.

FUNDING

This work was self-funded by the authors.

AUTHOR'S CONTRIBUTIONS

Doctors Larissa Campuzano and Lisa Arguello created the idea. Dr. Larissa Campuzano worked on the writing of the work. Dr. Evelio Legal and Dr. Marta Osorio on the final revision of the manuscript.

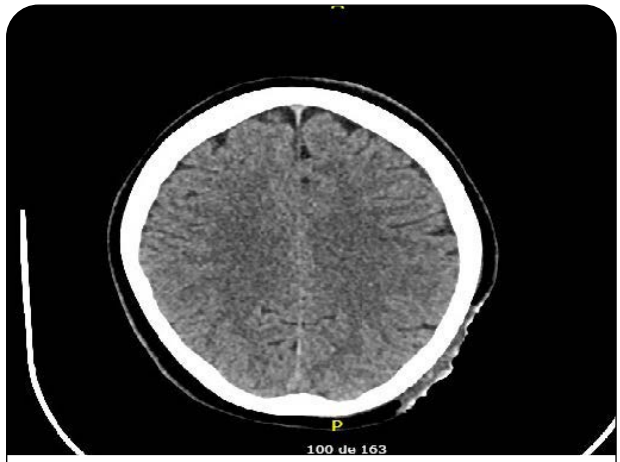

Figure 6. Postsurgical rutinary CT scan.

BIBLIOGRAPHIC REFERENCES

1. Eiger-Moscovich M, Eagle RC Jr, Shields CL, et al. Neoplasias sebáceas perioculares asociadas al síndrome de Muir-Torre: patrones de detección en la literatura y en la práctica clínica. Ocul Oncol Pathol. 2020;6(4):226-37.
2. Kibbi N, Worley B, Owen JL, et al. Carcinoma sebáceo: controversias y su evidencia para la práctica clínica. Arch Dermatol Res. 2020;312(1):25-31.
3. Sargen MR, Starrett GJ, Engels EA, Cahoon EK, Tucker MA, Goldstein AM. Sebaceous Carcinoma Epidemiology and Genetics: Emerging Concepts and Clinical Implications for Screening, Prevention, and Treatment. Clin Cancer Res. 2021;27(2):389-393.
4. Owen JL, Kibbi N, Worley B, Kelm RC, Wang JV, Barker CA, et al. Carcinoma sebáceo: guías de práctica clínica basadas en evidencia. Lancet Oncol. 2019;20:e699-e714.
5. Le DT, Durham JN, Smith KN, Wang H, Bartlett BR, Aulakh LK, et al. Mismatch repair deficiency predicts response of solid tumors to PD-1 blockade. Science. 2017;357:409-13.
6. Marcus L, Lemery SJ, Keegan P, Pazdur R. FDA Approval Summary: Pembrolizumab for the treatment of solid tumors with high microsatellite instability. Clin Cancer Res. 2019;25:3753-8.
7. Martín-Díaz, M. A., Mayor, M., Rubio, C., Hernández-Cano, N., González-Beato, M. J., & Casado, M. (2004). Carcinoma sebáceo extraocular de presentación atípica. Actas Dermo-Sifiliográficas, 95(9), 586-589.
8. Morgade, P., & Fraga, S. (2020). Carcinoma sebáceo extraocular. Revista argentina de cirugía, 112(4), 535-538.
9. Jiménez, R., Reichenberger, M. P., Valdés, F., Montes, R., Bustos, F., & Plass, I. (2023). Carcinoma sebáceo, 6 años de experiencia en el Instituto Nacional del Cáncer. Revista de cirugía, 75(4), 237-242.
10. Takagawa Y, Tamaki W, Suzuki S, Inaba K, Murakami N, Takahashi K, Igaki H, Nakayama Y, Shigematsu N, Itami J. Radiotherapy for localized sebaceous carcinoma of the eyelid: a retrospective analysis of 83 patients. J Radiat Res. 2019 Oct 23;60(5):622-629. doi: 10.1093/jrr/rrz046. PMID: 31271440; PMCID: PMC6805970.