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CASE REPORT

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Lacrimal sac squamous cell carcinoma: From resection to prosthetic rehabilitation. A case report

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Key Clinical Message

There is a lack of consensus and evidence on treatment strategies for lacrimal sac carcinomas. Wide en bloc surgical resection with farther prosthetic rehabilitation could be the treatment option in certain cases.

Abstract

Malignant epithelial lacrimal sac tumors are rare cancers with high recurrence rates. Diagnosis of these tumors is often delayed as they are confused with chronic dacryocystitis. There is a lack of consensus and evidence on standard treatment strategies for advanced lacrimal sac carcinomas. A case of advanced lacrimal sac squamous cell carcinoma treated with wide en bloc margin-negative surgical resection with further prosthetic rehabilitation without adjuvant therapy and 38 months of recurrence-free postoperative follow-up is presented.

KEYWORDS

carcinoma en bloc resection, facial prosthetic rehabilitation, lacrimal sac carcinoma, magnabar fixation, squamous cell carcinoma

1 | INTRODUCTION

Malignant epithelial lacrimal sac tumors are rare cancers with high recurrence rates.[1–6] A total of 539 cases were reported in the literature from 1960 to 2019, among which squamous cell carcinoma, with 296 cases, was the most commonly described.[4]

The clinical presentation of lacrimal sac squamous cell carcinoma (LSSCC) resembles that of chronic dacryocystitis, which does not disturb until the appearance of specific symptoms, such as blood-stained tearsora, palpable lump, or a progressive mass in the area of the lacrimal sac/nasolacrimal duct.[3,4,7] Thus, diagnosis is often delayed in LSSCC, and in some instances, the diagnosis is missed even during routine dacryocystorhinostomy. In these cases, the tumor can grow into the adjacent sinuses and the nasal cavity and cause significant morbidity.[3] Fewer than 15% of cases of LSSCC are diagnosed within 2 months, and treatment is initiated within 12 months in 72% of patients.[1] MRI or computed tomography (CT) scan of the orbit or paranasal sinuses is the preferred imaging modality to diagnose lacrimal sac tumors.[6]

Complete surgical excision followed by radiotherapy is the preferred modality of management, and only 18% require orbital exenteration.[5] Wide surgical en bloc resection of lacrimal sac tumors with maxillectomy provided

good results in reducing the incidence of recurrence of the disease. Orbit exenteration, resection of the paranasal sinuses, or lymph node dissection is performed in certain advanced cases.[7–9] Lymph node status was found to be a key factor for prognosis.[10]

Because of the anatomic location of the lacrimal sac and nasolacrimal duct and their proximity to the orbital soft tissue, the maxilla and paranasal sinuses a multidisciplinary surgical approach is often optimal. Some clinicians avoid attempting globe-sparing surgery because of concerns about a higher risk of local recurrence if the eye is spared and because of concerns about ocular damage from radiation therapy (RT).[3]

A case of advanced LSSCC treated with wide en bloc margin-negative surgical resection with further prosthetic rehabilitation without adjuvant therapy and 38 months of recurrence-free postoperative follow-up is presented.

2 | CASE HISTORY/ EXAMINATION

A 30-year-old man was admitted to the Department of ENT and Maxillofacial Surgery with complaints of a slightly painful progressive mass in the area of the lacrimal sac. He presented a 1-year history of unilateral epiphora in the right eye following acute ipsilateral inflammation (dacryocystitis) of the medial canthal region. The patient was treated with short-term systemic antibiotic therapy prescribed by the family doctor and then referred for specialized evaluation. On clinical examination, right-side extensive reddish bulging mass was revealed on the medial cantus region with lower and upper eyelid involvement (Figure 1). On palpation, the mass was firm and moderately painful. An incisional biopsy of the mass was performed under local anesthesia and squamous cell carcinoma of the lacrimal sac was disclosed.

2.1 | Differential diagnosis, investigations, and treatment

Contrast head and neck CT scan examination revealed a 2.5 × 1.6 × 3.2 cm neoplastic formation with involvement of the lacrimal sac/duct, the medial part of the right orbit, the medial rectus muscle with very close adjustment of the eyeball without visible borders (Figure 2A). There was orbit inferior-medial bone wall and lacrimal bone resorption and invasion of neoplastic formation to the nasal cavity close to the medial nasal concha and maxillary sinus (Figure 2B). Right-side carotid group lymph nodes slight enlargement was revealed with a size of 1.3 × 0.9 cm and submandibular lymph nodes with a size of 1.1 × 0.8 cm. The orbits contrast CT scan (DLP-1530.0 mg) did not reveal lesion invasion into the eyeball.

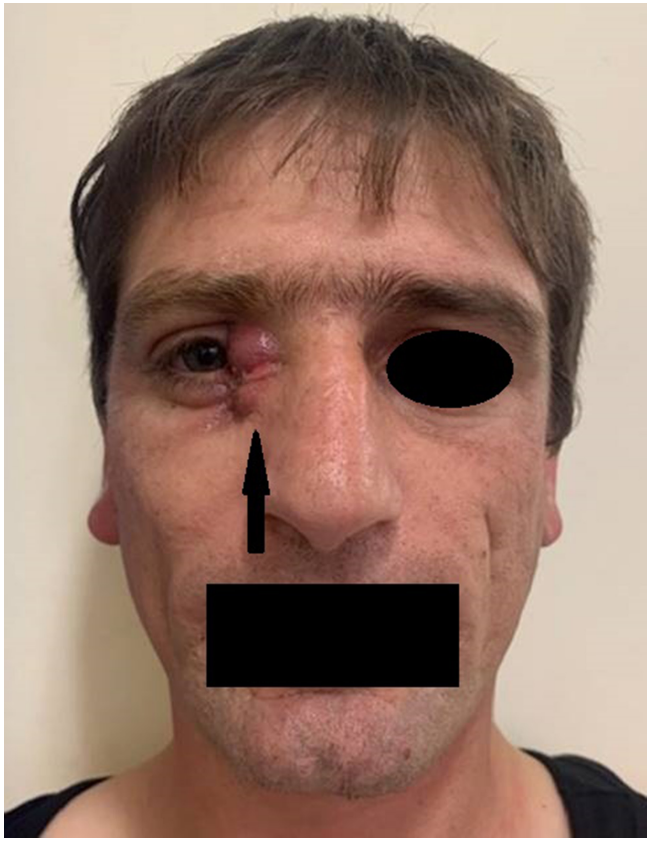

FIGURE 1 External view of the patient's face—extensive reddish bulging mass on the medial cantus region with the lower and upper eyelids involvement.

Fine needle aspiration of regional sentinel lymph node did not reveal metastatic involvement. Right-side wide en bloc resection with orbit exenteration including the ethmoid, lateral nasal bone, lacrimal sac and duct, and maxillary sinus upper wall without neck lymphodissection was performed (Figure 3A,B). The frontal musculocutaneous flap was used for lateral nasal defect closure (Figure 3C).

The histological examination revealed cohesive atypical squamous cell tumor complexes composed of cells with moderately atypical nuclei and eosinophilic cytoplasm. Few foci of keratinization and necrosis were found. There were mitoses, including atypical ones. A moderately expressed mixed inflammatory infiltrate with a predominance of eosinophils was observed in the stroma (Figure 4). The tumor grew into the adjacent soft tissues and upper bone. There were no lymphovascular or perineural embolisms. The cutting edges were ablastic. The final histological diagnosis was lacrimal sac/duct squamous cell carcinoma G2 pT4a pNx LO VO PnO RO.

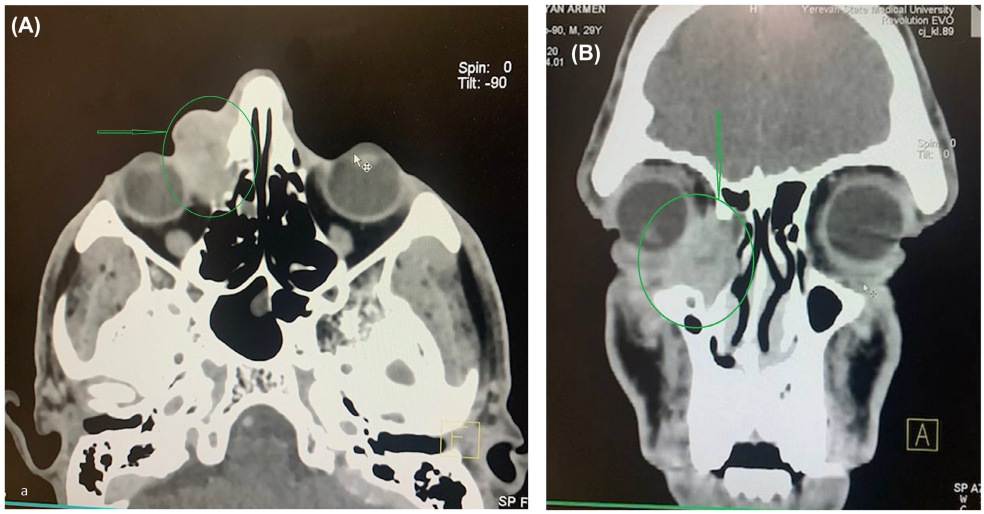

FIGURE 2 Post-contrast-enhanced CT shows tumor extension from the lacrimal sac/duct into the medial canthus, medial rectus muscle, ethmoid, nasal cavity, medial concha, and maxillary sinus: axial view (A), coronal view (B).

FIGURE 3 Operating field after en bloc resection (A), macroscopic view of resected block from outside (B), lateral nasal defect closure with frontal musculocutaneous flap (C).

The post-exenteration wound healing was performed under iodoform gauze. It takes approximately 1 month for secondary intention wound healing under gauze.

During the first postoperative year, quarterly control head and neck contrast CT scan examinations were performed. No signs of recurrence were observed (Figure 5).

Therefore, prosthetic rehabilitation was planned for the patient.

Under local anesthesia, two dental implants were installed in the lateral zygoma (Figure 6). After 4 months, healing caps were adopted through the small skin incision.

A silicone implant-supported epithesis was constructed for the patient with a right zygo-orbital defect for a total esthetic rehabilitation. The magnabar retention system was selected as the method of choice for the patient due to the depth and volume of the defect. The main magnet was situated on a casted framework bar at the lateral perimeter of the prosthetic field. The ocular was made of crystal clear acrylic (non-free-monomer methyl-met-acrylate, class 3 of ANNEX IX classification) in the scleral portion, and the iris was painted in the conventional methods of iris painting used in ocular prosthetics to achieve a good esthetic result in comparison with the left eye (Figure 7A,B).

Silicone material of the orbital epithesis was made of a VTR platinum silicone of Technovent M511 with 25 Shore. Magnet keepers for the matrix magnet were made of a self-cure Vilacryl methacrylate. The silicone coloring system used in this epithesis was intrinsically pigmented silicone layering with negative painting of the stone cast.

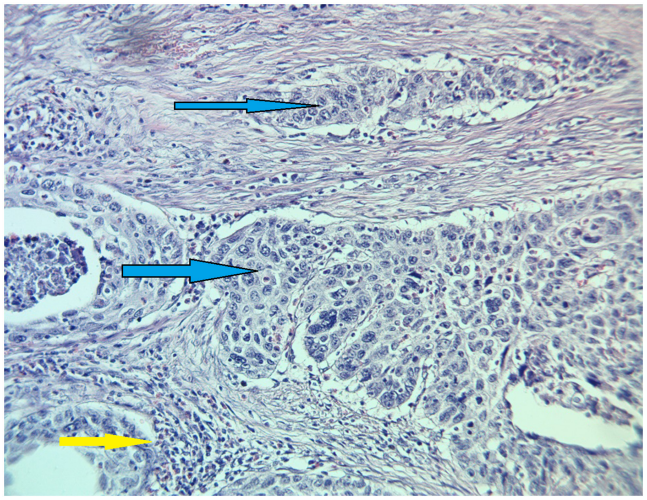

FIGURE 4 Nests of squamous cell carcinoma (blue arrow) with surrounding inflammation (yellow arrow) × 40, H@E.

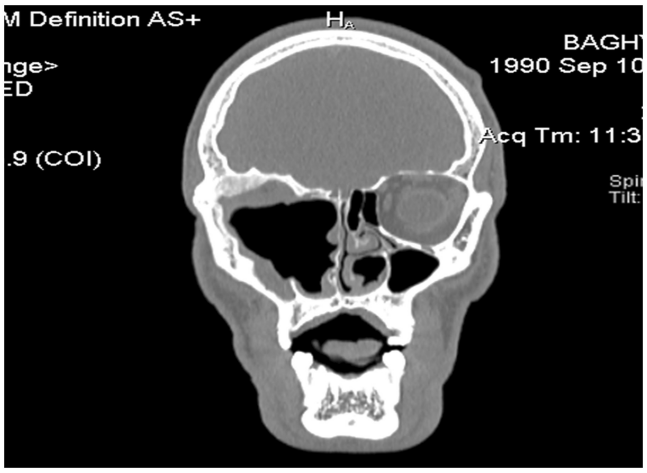

FIGURE 5 Coronal CT scan of the patient after 1 year follow-up.

3 | OUTCOME AND FOLLOW-UP

The 38-month postoperative follow-up did not reveal signs of recurrence in the present case.

4 | DISCUSSION

Primary epithelial malignancies of the lacrimal apparatus, comprising the lacrimal gland, duct, and sac, are extraordinarily and uncommon rare tumors with significant recurrence rates.[1–4,11–14] As noted by Singh and Ali[5] among primary malignant epithelial tumors, squamous cell carcinomas were the most common (61%), followed by transitional cell carcinomas (15%), and mucoepidermoid carcinoma (7%). Because of their rarity, no extensive clinical data on their management and prognosis exist.[7]

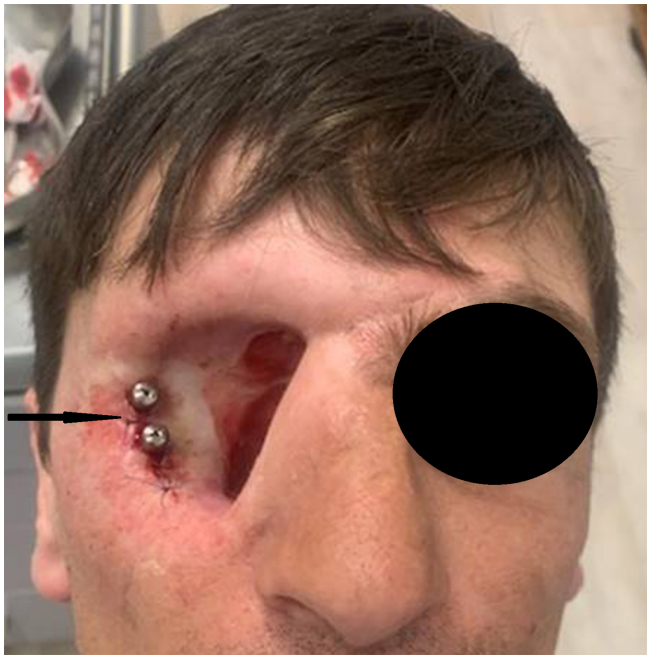

FIGURE 6 Two dental implants were installed in the lateral zygoma.

Mortality rates for malignant tumors depend on tumor stage and type, and the mean rate is 38%.[1] Lacrimal drainage system carcinoma is a disease of the middle-aged and elderly population. Studies have reported that the malignant lacrimal sac tumors often occur in the fifth decade with a slight male predominance.[4,10,13,15] Symptoms of lacrimal sac carcinoma are in many ways similar to those of a benign disease of the lacrimal system. Due to these tumors rarity, there is often a long-term lack of accurate diagnosis, due to the fact that they are confused with dacryocystitis.[1,4,6,7,12,13] Swelling in the medial canthal region, epiphora, and pain are the most common presenting features.[6,13] LSSCC can involve the lacrimal sac and grow through the nasolacrimal duct with invasion to peripheral organs and structures.[7] These patients are often referred later after the revealing of a malignant tumor on biopsies of the lacrimal sac taken when dacryocystitis recurs.[1,16]

In the present case, the 30-year-old male patient had a 1 year history of right eye epiphora and redness and swelling in the medial cantus area for the preceding 5 months. Dacryocystitis was diagnosed by the family doctor and antibiotics were prescribed. After 3 months of unsuccessful conservative treatment, the patient was directed to hospital treatment where the deep incisional biopsy was performed and a diagnosis of LSSCC was clarified.

Thorough clinical investigation and computed tomographic-dacryocystography, CT scan of the orbit, or magnetic resonance dacryocystography can help in diagnosing a lacrimal sac tumor. Imaging is essential for identifying the location, size, and extent of the lesion,

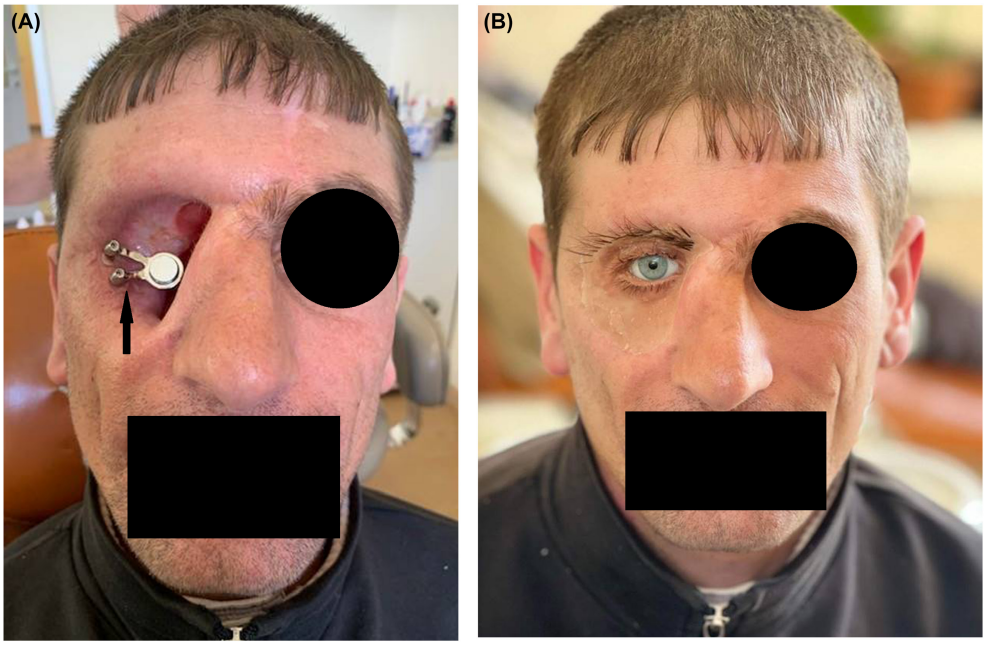

FIGURE 7 External view of the patient's face: magnabar retention system fixed on implants (A), a silicone implant-supported epithesis fixed on a magnabar retention system (B).

assessing the disease severity, and differentiating tumors from inflammatory and infectious lesions.[17] CT scans of the orbit or paranasal sinuses with axial, coronal, or sagittal images are used to diagnose lacrimal sac tumors and to assess osteolytic changes as well as the invaded surrounding tissues.[3,6,11]

Kumar et al. recommend performing a thin-section (1.25-mm) CT with contrast as the first-line imaging study to evaluate malignant lacrimal sac and nasolacrimal duct tumors at the time of initial staging. MR imaging can be performed if CT cannot distinguish sinonasal tumor extension from postobstructive secretions.[11]

In the present study. head, neck, and chest CT scans with contrast and isolated CT scans of orbits were performed.

Correct diagnosis and appropriate therapy require a multidisciplinary management approach. First and foremost in the treatment of these malignant epithelial tumors is complete surgical removal with wide excision.[1,12]

However, there is no standard surgical treatment strategy. Multidisciplinary therapy, including surgery, chemotherapy, and radiotherapy, is the primary treatment modality.[3,4] Wide surgical en bloc resection of lacrimal sac tumors with medial maxillectomy or total maxillectomy is favored with good success rates for local disease control.[7,9] Orbit exenteration, resection of the paranasal sinuses, or lymph node dissection is performed in certain advanced cases.[8,18] Song et al.[10] reported that the outcomes of comprehensive treatment were quite encouraging, and the 5-year overall survival rate and 5-year progression-free survival rate were 87.6% ± 4.8% and 76.3% ± 6.4%, respectively.

The combined sinus–orbit approach is an effective method of managing lacrimal sac tumors to achieve optimal tumor clearance from the orbit and nasal cavity.[9]

Decreased recurrence rates were observed in patients who underwent lateral rhinostomy and wide excision compared to those without rhinostomy.[6] Aggressive malignant lesions may require the removal of the entire lacrimal drainage system, including the canaliculi, lacrimal sac, and entire nasolacrimal duct with lateral rhinotomy.[5,6] Orbital exenteration and resection of the paranasal sinus may be needed for extensive primary or secondary malignant lesions.[6,8,12,19] The balance of surgical radicality and preserving quality of life is similar to trade off thinking for these cases.[14] In the present case, the neoplastic lesion was invaded the medial rectus orbital muscle, ethmoid, nasal cavity, medial concha and maxillary sinus. Wide en bloc surgical removal could be more life-lengthening for this case than organ-preserving operation tactics and RT. Notably, lymph node status was a key factor in determining outcomes.[1,7]

RT has been considered an alternative to surgery, but there is no consensus on its use for advanced lacrimal sac carcinoma. Song et al.[7] in their study of 17 cases concluded that RT alone achieved excellent long-term clinical outcomes and could be a viable treatment option for the patients who refused surgery or had unresectable tumors. They have described the detailed methodology of the provided RT. The treatment was planning on a three-dimensional CT image-based planning system, where the eyeballs, lens, optic nerves, and optic chiasma were outlined. The gross tumor volume was defined and delineation of two clinical target volumes for the primary

tumor and higher and lower tumor burden, which should be associated with a higher and a lower dose prescription were performed. Patients were irradiated using intensity-modulated radiation therapy (IMRT) or conventional three-dimensional-conformed RT during the entire RT treatment. A6 MV linear accelerator or electron-beam irradiation to the primary tumor was performed and a total dose of 6600–7000 cGy was prescribed. The nine irradiation beams were angled in IMRT to avoid the cornea and retina radiation damage.

There is also no high-quality evidence on the use of chemotherapy in advanced lacrimal sac carcinoma to date. In sum, there is a lack of consensus and evidence on standard treatment strategies for advanced lacrimal sac carcinoma.[14] Adjuvant treatment modalities include external beam RT, local RT (plaque brachytherapy), proton therapy, chemotherapy (CHOP regimen) or immunotherapy.[6,20] Ashok Kumar et al.[21] in their study provided the retrospective cohort analysis of epithelial lacrimal gland tumors type of surgery and various treatment modalities from the National Cancer Database. Patients were divided into eight groups, based on the combinations of adjuvant or neoadjuvant chemotherapy or RT used. Surgery with adjuvant RT was the most frequently used treatment modality in described study. The authors was found no difference in outcome between destructive and orbit sparing procedures nor the use of any form of chemotherapy or RT.

Recurrence and mortality rates for lacrimal sac tumors vary from case to case.[3,4,6–10,16,18,20] In the present study, the patient did not receive any postoperative adjuvant treatment and no recurrence was revealed during 38 months of follow-up.

To our knowledge, this is the first report of a case of facial prosthetic rehabilitation after advanced LSSCC wide en bloc resection without adjuvant therapy.

5 | CONCLUSION

Malignant epithelial lacrimal sac tumors are rare cancers with high recurrence rates. Diagnosis of these tumors is often delayed because they are confused with dacryocystitis. Multidisciplinary therapy, including surgery, chemotherapy, and radiotherapy, is the primary treatment modality. Wide surgical en bloc resection with orbit exenteration is recommended as a surgical tactic for advanced cases. Facial prosthetic rehabilitation could be an effective method in the recurrence free postoperative period.

AUTHOR CONTRIBUTIONS

Anna Poghosyan: Project administration; supervision; writing – review and editing. **Armine Gharakeshishyan:** Data curation; methodology; visualization. **Martin Misakyan:** Conceptualization; formal analysis; resources. **Davit Minasyan:** Methodology; software. **Parandzem Khachatryan:** Data curation; investigation. **Karen Mashinyan:** Formal analysis; methodology; writing – original draft. **Sergo Hovhannisyan:** Data curation; investigation; visualization. **Artavazd Kharazyan:** Conceptualization; methodology; validation; visualization.

CONFLICT OF INTEREST STATEMENT

The authors have no conflicts of interest to declare.

FUNDING INFORMATION

None.

DATA AVAILABILITY STATEMENT

Data sharing not applicable to this article as no datasets were generated or analyzed during the current study.

ETHICS STATEMENT

We confirm that explicit written consent to publish the results has been received from the described patient.

CONSENT

Written informed consent was obtained from the patients for the publication of this case report and any accompanying images. A copy of the written consent is available for review to the editor-in-chief of this journal.

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