

# A Navajo Patient with Morbihan's Disease: Insight into Oculoplastic Treatment of a Rare Disease

Lacey R. Pflibsen, MD\*  
 Ashley L. Howarth, MD\*  
 Ana Meza Rochin, BS†  
 Tim Decapite, MD‡  
 William J. Casey III, MD\*  
 Lisa A. Mansueto, MD\*§

**Summary:** Morbihan's disease is classically defined as primarily a Caucasian disease of chronic, recurrent erythema and nonpitting edema of the middle and upper thirds of the face. The disease is often difficult to diagnose and challenging to treat. Medical management is the primary treatment modality; however, if there is progression of the disease that causes visual impairment, surgical debulking is often required. We present the first reported case of Morbihan's disease in a Navajo patient in conjunction with a review of the literature on surgical management of this challenging disease. (*Plast Reconstr Surg Glob Open* 2020;8:e3090; doi: 10.1097/GOX.0000000000003090; Published online 17 September 2020.)

**M**orbihan's disease, also known as rosacea lymphedema or solid persistent facial edema, is a rare disease of chronic, recurrent erythema and nonpitting edema that primarily affects the middle and upper thirds of the face.<sup>1,2</sup> "Morbihan" refers to the district in France, where the disease's classic findings were described by Robert Degos in 1957.<sup>3</sup> It is unclear whether this disease is its own entity or rather an end-stage complication of rosacea.<sup>2</sup> The diagnosis is based on clinical presentation, histological evaluation, and exclusion.<sup>2</sup> Histological findings vary greatly in the literature, though they typically include dilated blood vessels, perifollicular fibrosis, and perivascular and perifollicular infiltration of lymphocytes.<sup>1,2,4</sup> It is primarily treated medically with antibiotics or steroids or isotretinoin therapy, or with a combination of these.<sup>1</sup> Morbihan's disease, especially cases resistant to medical treatment with periocular involvement, can lead to cosmetic disfigurement and visual obstruction, for which surgical intervention is warranted.<sup>1,5–7</sup>

Morbihan's disease was classically defined as a Caucasian disease.<sup>1,3</sup> Morbihan's disease has only been reported in a few non-white patients, including those of

Asian, Hispanic, Black, and Asian Indian descent.<sup>1,3,8</sup> We present the first reported case of Morbihan's disease in a Navajo man and a review of the surgical management of this challenging disease.

## CASE

A 55-year-old Navajo man, who resides on the Navajo Reservation, presented with a 1-year history of left upper lid (LUL) swelling (*Fig. 1*). His medical history was significant for a recent diagnosis of rosacea and type 2 diabetes mellitus. Oral doxycycline (100 mg twice daily) and topical metronidazole (1% gel applied daily) had been prescribed for 6 months before initial surgical consultation with no significant effect. A maxillofacial CT scan was performed, demonstrating preseptal thickening and inflammation of the LUL. A punch biopsy revealed chronic inflammation, some granulomas, and histiocytes. Microbiologic workup was negative. These findings were consistent with orofacial granulomatosis or granulomatous rosacea. Ocular examination showed severe eyelid edema with induration, margin to reflex distance of –1 mm, and levator palpebrae superioris excursion of 12 mm. Owing to the significant visual impairment, a LUL debulking of the excess dermatochalasis, edematous and fibrotic orbicularis oculi muscle, and thickened septum of the upper lid was performed via a standard upper lid blepharoplasty approach: A 1.2 cm (vertical height) by 2.8 cm (width) section of skin was resected. Pathological examination showed findings similar to the previous punch biopsy of chronic inflammation, some granulomas, and histiocytes. His visual obstruction significantly improved and there was no lagophthalmos. However, he returned 3 years later with contralateral swelling and erythema of his right upper lid (*Fig. 2*). Right upper lid debulking was performed through an upper blepharoplasty incision, and edematous and fibrotic orbicularis oculi muscle and thickened septum were again resected.

From the \*Division of Plastic Surgery, Department of Surgery, Mayo Clinic Arizona, Phoenix, Ariz.; †University of Arizona College of Medicine, Phoenix, Ariz.; ‡Department of Dermatology, Tuba City Regional Health Care Corporation, Tuba City, Ariz.; and §Orbit and Oculofacial Plastic Surgery, Department of Ophthalmology, Ophthalmic Surgeons & Physicians, Ltd., Tempe, Ariz.

Received for publication June 21, 2020; accepted July 15, 2020.

Presented at the Mountain West Society of Plastic Surgeons, February 2020, Snowmass Village, Colo.

Copyright © 2020 The Authors. Published by Wolters Kluwer Health, Inc. on behalf of The American Society of Plastic Surgeons. This is an open-access article distributed under the terms of the [Creative Commons Attribution-Non Commercial-No Derivatives License 4.0 \(CCBY-NC-ND\)](#), where it is permissible to download and share the work provided it is properly cited. The work cannot be changed in any way or used commercially without permission from the journal.

DOI: 10.1097/GOX.0000000000003090

**Disclosure:** The authors have no financial interest to declare in relation to the content of this article.



**Fig. 1.** Preoperative photograph of a 55-year-old Navajo male patient showing left upper eyelid swelling, erythema, and visual obstruction.

A 1.4cm (vertical height) by 2.8cm (width) of tissue was resected. There was no functional disturbance of the eyelid after surgery. Pathological examination showed the same findings. At this point, the diagnosis of Morbihan's disease was made due to his symptoms of the upper and middle thirds of the face, prior specimen histologic findings, and exclusion of other possible differential diagnoses such as Melkersson–Rosenthal syndrome, systemic lupus erythematosus, dermatomyositis, sarcoidosis, chronic actinic dermatitis, and contact dermatitis.<sup>3</sup> Trials of methotrexate and adalimumab failed to result in any improvement. Oral prednisone was initiated with improvement of cutaneous symptoms; however, his diabetes became uncontrolled and oral prednisone was discontinued. One year later, he returned with recurrent bilateral upper and lower eyelid edema. Additional surgical debulking and injection of triamcinolone (40mg/mL) were performed: 1mL into each upper eyelid. His symptoms improved with the addition of the steroid injections, as his eyelids softened and became less edematous (Fig. 3). He was subsequently lost to follow-up. When he returned 2 years later, he had recurrence of symptoms in bilateral upper and lower eyelids. A second opinion from an outside oculofacial plastic surgeon was sought, and the diagnosis of Morbihan's disease was confirmed, with no suggested deviation from the proposed medical or surgical management. Dermatology initiated isotretinoin orally, but despite this the patient had recurrence of edema with associated visual impairment bilaterally 1 year after its initiation (Fig. 4). The patient did not desire repeated surgical debulking and has been lost to follow up.



**Fig. 2.** Image of the same patient showing the involvement of contralateral upper and lower lid with visual impairment, 3 years after initial debulking.

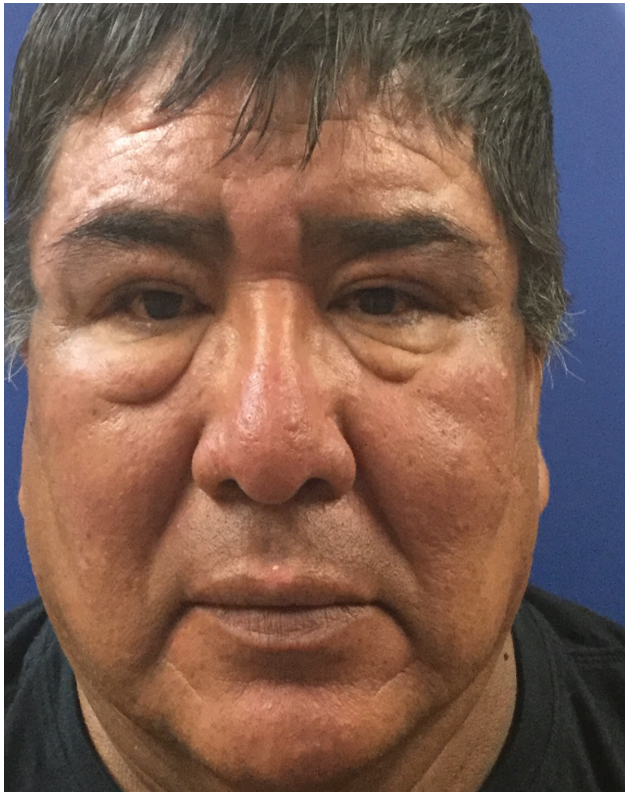
## DISCUSSION

There is a paucity of data on the surgical management of Morbihan's disease. Surgery is reserved for failure of primary medical therapy without significant improvement in symptoms of periocular involvement such as visual obstruction or ectropion.<sup>5–7,9</sup> Most reported cases have muscular and septal involvement with fibrosis and edema.<sup>5,9</sup> For upper eyelids, most surgeons report using the standard blepharoplasty incision.<sup>5,7</sup> For lower lids, most utilize a subciliary incision and excise involved portions of the orbicularis oculi muscle and orbital septum.<sup>5,7,9</sup> Our patient had significant periocular involvement with visual obstruction and involvement of the orbicularis oculi muscle and orbital septum, which were resected. In one case report, surgeons utilized CO<sub>2</sub> laser cautery during surgery with the debulking via blepharoplasty technique.<sup>5</sup> The authors hypothesized that the laser might result in better lymphatic vessel occlusion possibly decreasing relapse rate.<sup>5</sup> However, no further studies were found evaluating this proposed techniques' efficacy.

Another described surgical technique resects all involved tissues. Méndez-Fernández et al<sup>6</sup> surgically debulked the affected tissues en bloc, including the skin, and placed a split thickness skin graft on the resulting defect. This led to significant ectropion, ultimately requiring eventual full thickness skin grafts and bilateral canthopexies.<sup>6</sup>

In summary, there is no gold standard for the surgical management of this rare and complex disease. In our





**Fig. 3.** Three months postoperative photograph of the patient after the bilateral upper lid surgery and triamcinolone injections.

experience, this disease has preseptal fibrosis and inflammation involving the orbicularis oculi muscle and orbital septum. Utilizing a blepharoplasty incision for debulking allows for better cosmesis and avoids skin grafting, which can lead to ectropion or scleral show.<sup>6</sup> Our patient had recurrence of symptoms after primary disease debulking, even with adjunct medical treatment. In affected patients, additional debulking procedures utilizing previous incisions are possible, as seen in repeat cosmetic blepharoplasties. Although there appears to be recurrence of symptoms in many of these patients, it is our opinion that surgical debulking utilizing a blepharoplasty incision and local steroid injections is the preferred approach for this disease, as it allows for better cosmesis and repeat surgical debulking.

We present the first reported Navajo patient diagnosed with Morbihan's disease. This disease is primarily described in white patients, and limited studies have discussed this disease in other ethnicities. In a recent systematic review by Boparai et al, there have been 11 Asian, 2 Hispanic, 1 Black, and 1 Asian Indian cases reported in the literature to date.<sup>1,3</sup> It is important to document this diagnosis in a Navajo patient, as it is rare in non-white patients, and to add this case to the growing body of literature about the epidemiology, diagnosis, and treatment of this disease.

### CONCLUSIONS

This is the first report of Morbihan's disease in a Navajo patient. The authors recommend utilizing a



**Fig. 4.** Image of the patient showing bilateral recurrence of edema and erythema.

traditional upper lid or subciliary lower lid blepharoplasty incision for surgical debulking and local steroid injection when surgical intervention is warranted based on our experience, though no gold standard has been formally reported.

**Lisa A. Mansueto, MD**

3200 S. Country Club Way  
Tempe, AZ 85282

E-mail: [drmansueto@osandp.com](mailto:drmansueto@osandp.com)

### PATIENT CONSENT

*The patient provided written consent for the use of his image.*

### REFERENCES

1. Boparai RS, Levin AM, Lelli GJ Jr. Morbihan disease treatment: two case reports and a systematic literature review. *Ophthalmic Plast Reconstr Surg*. 2019;35:126–132.
2. Smith LA, Cohen DE. Successful long-term use of oral isotretinoin for the management of Morbihan disease: a case series report and review of the literature. *Arch Dermatol*. 2012;148:1395–1398.
3. Veraldi S, Persico MC, Francia C. Morbihan syndrome. *Indian Dermatol Online J*. 2013;4:122–124.
4. Morales-Burgos A, Alvarez Del Manzano G, Sánchez JL, et al. Persistent eyelid swelling in a patient with rosacea. *PR Health Sci J*. 2009;28:80–82.
5. Bechara FG, Jansen T, Losch R, et al. Morbihan's disease: treatment with CO<sub>2</sub> laser blepharoplasty. *J Dermatol*. 2004;31:113–115.

6. Méndez-Fernández MA. Surgical treatment of solid facial edema: when everything else fails. *Ann Plast Surg*. 1997;39:620–623.
7. Carruth BP, Meyer DR, Wladis EJ, et al. Extreme eyelid lymphedema associated with rosacea (Morbihan disease): case series, literature review, and therapeutic considerations. *Ophthalmic Plast Reconstr Surg*. 2017;33(3S Suppl 1):S34–S38.
8. Bhopal R. Glossary of terms relating to ethnicity and race: for reflection and debate. *J Epidemiol Community Health*. 2004;58:441–445.
9. Bernardini FP, Kersten RC, Khouri LM, et al. Chronic eyelid lymphedema and acne rosacea. Report of two cases. *Ophthalmology*. 2000;107:2220–2223.