

# Microcystic Adnexal Carcinoma: Reconstruction of a Large Centrofacial Defect

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**Summary:** We report a rare case of a large facial microcystic adnexal carcinoma in an elderly patient who underwent several rounds of excision. The patient was left with a large facial defect and remaining positive margins. The decision was made to stop further excision and proceed with reconstructive surgery. We show that the patient's function and quality of life were not impeded despite reconstruction in light of positive margins for tumor. We believe that this case will draw the surgeon's attention to the possibility of palliative reconstruction in the treatment of a patient with large debilitating facial defects after microcystic adnexal carcinoma excision. (*Plast Reconstr Surg Glob Open* 2014;2:e254; doi: 10.1097/GOX.000000000000195; Published online 21 October 2014.)

**M**icrocystic adnexal carcinoma (MAC) is a rare, malignant tumor of the skin that is commonly classified as a low-grade sweat gland carcinoma. Goldstein et al<sup>1</sup> first described the tumor in 1982. The tumor arises in the skin adnexa from a pluripotent adnexal keratinocyte. Histologically, it is well differentiated and appears as small, keratinized cysts with well-defined ducts.<sup>2</sup>

The Surveillance, Epidemiology and End Results database was able to identify a total of 273 reported cases from 1973 to 2004.<sup>3</sup> Yu et al<sup>3</sup> concluded that MAC is a locally invasive tumor that rarely metastasizes and has a predilection for the head and neck skin. The clinical presentation is typically a firm, flesh-colored nodule or plaque.<sup>4</sup>

Kirkland et al<sup>5</sup> reported that the tumor often requires extensive surgical resection. Outcome studies

of tumors treated with Mohs micrographic surgery (MMS) show that MMS allows for tissue conservation and an increased likelihood of long-term cure with one study reporting a 2-year success rate of 89.7%.<sup>4,6</sup> Limited data exist regarding the role of radiation therapy, but a recent study showed promising outcomes.<sup>7</sup>

The purpose of our case report is to add to the limited treatment experience of this rare tumor.

## CASE REPORT

An 85-year-old otherwise healthy woman was referred from a major academic center department of dermatology after undergoing 12 stages of MMS. She originally presented to dermatology with a subcentimeter pearly papule of the left nasal tip (Fig. 1). Examination of the initial specimen revealed a typical infiltrating basal cell carcinoma of the deep reticular dermis. A second layer of tissue was obtained and focal areas of basal cell carcinoma were present. Interestingly, there were also many areas of syringomatous-like structures in the dermis that were characterized by small ductules containing abundant basophilic material and an associated host lymphocytic response. Initially, a diagnosis of an unusual morpheaform basal cell carcinoma was made. However, on further inspection, the syringomatous-like structures invaded deeply into the reticular dermis, subcutaneous fat, and underlying nasalis musculature. Given the overall histologic

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**Fig. 1.** Preoperative photograph showing the original basal cell carcinoma.

appearance coupled with the dermal response, MAC was added to the differential diagnosis. On further clinical examination, a subcentimeter flesh-colored plaque was visualized on the nasal dorsal surface. Examination of successive specimens revealed continued proliferation of the syringomatous-type lesions. There was no evidence of perineural invasion. The patient and her family were notified of a second cutaneous tumor of the face that appeared to be deeply invasive and multifocal. The patient continued to undergo a total of 12 stages of MMS in attempts to attain tumor-free tissue planes. After the twelfth stage, a 12×8cm soft-tissue defect of the nose, glabella, and bilateral cheeks was present as well as extensive positive multifocal margins. The specimens were reviewed by multiple dermatopathologists both within and outside the institution, all of whom agreed with a diagnosis of MAC. The patient and her family then elected to terminate further excision despite positive margins. They were informed that the exact biological potential of the remaining disease could not be predicted and that further excision might be necessary if the tumor invaded a vital structure.

The patient was then referred to plastic surgery with a 12×8cm soft-tissue defect involving the nose, glabella, and bilateral cheeks (Fig. 2). Multiple reconstructive options were discussed. Coverage of the nasal defect was obtained with full-thickness skin grafts from bilateral supraclavicular fossae and the midline neck. Closure of the forehead and cheek defects was obtained with an A:T advancement flap and bilateral Mustarde cheek flaps, respectively. The patient and her family elected for this more cosmetically appealing option understanding that flap coverage might obscure future tumor surveillance. Successful coverage was achieved (Fig. 3). At a 3-year follow-up, the patient was satisfied with her functional and cosmetic outcome. No clinical evidence of metastasis or persistent tumor was identified.



**Fig. 2.** Preoperative photograph showing the large centrofacial defect after MMS.

### SUMMARY

Given the rarity of this tumor, we faced a dilemma regarding wound closure in light of extensive positive margins. Although the tumor is known to be locally aggressive, invading skin, muscle, vascular adventitia, nerves, perichondrium, and periosteum, to date there are only 6 reported cases of metastasis.<sup>8,9</sup> There is one reported death from this tumor, giving a death rate of 0.4%.<sup>10</sup> A published case report showed that observation despite positive margins may be a reasonable option.<sup>11</sup> Eisen and Zloty<sup>11</sup> reported a case where there were extensive margins beyond visible borders shown by multiple biopsies that would require extensive facial excision for complete resection. The authors elected against further excision and chose to observe the patient. At a 2-year follow-up, there were no signs of metastasis or persistent local disease.

Radiation therapy is another studied treatment modality. The largest published case series regarding the role of adjuvant radiation therapy in the treatment of MAC concluded that doses of 50 Gy or greater should be considered when further excision is not feasible. All



**Fig. 3.** Postoperative photograph at 3-year follow-up showing adequate reconstruction and functional outcome despite positive margins.

of the patients in their series underwent local excision and in those who had positive margins, radiation was performed rather than re-excision. Ten of 11 patients had positive margins that did not recur after radiation.<sup>12</sup>

Chemotherapy, as a treatment for MAC, has been reported once in the literature. A single patient received a course of cisplatin and 5-Fluorouracil after extensive surgical resection and radiation therapy. Tumor persisted requiring further treatment with surgery and radiation therapy.<sup>13</sup>

Our experience reveals that it can be challenging to obtain negative margins without disfigurement or functional deficits. Plastic surgeons are often faced with the task of wound coverage while attempting to optimize aesthetic and functional outcomes. An ethical dilemma existed regarding reconstruction of our patient's facial defect despite positive margins. Given our patient's age and desire to minimize further disfigurement, she elected against further excision, which in her case we felt was reasonable given this tumor's low risk of metastasis and death. In this case, reconstruction in the face of positive margins did not impede the quality of life or cause further morbidity or mortality in this elderly patient. To our knowledge, this is the second report to document reconstruction

in light of positive margins. Of course, it is imperative to attempt to obtain complete resection of the tumor as excision continues to be the mainstay treatment. However, when other circumstances prevail, radiation therapy and/or observation may be considered.

## CONCLUSIONS

This case highlights the possibility of palliative reconstruction when faced with disfigurement after extensive excision of MAC of the face.

## PATIENT CONSENT

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

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