

Priyenka Khatiwada BS¹, Jane Gay MD², and Douglas J. Grider MD^{1,2}

¹Virginia Tech Carilion School of Medicine (VTC SOM)

²Carilion Clinic Dermatology and Mohs Surgery



Background

- Microcystic adnexal carcinoma (MAC) is a rare, malignant sweat gland tumor that aggressively invades deep into local tissue. It is associated with a high rate of misdiagnosis.¹
- It is most often reported as a cutaneous lesion on the head and neck.¹
- Treating MAC with standard wide local excision has a high rate of recurrence (60%), and treatment with Mohs surgery is associated with higher cure rates.² Facial lesions requiring multiple re-excisions can be disfiguring.²
- Histopathologically, MAC is characterized by small keratocysts in the upper dermis with a deep infiltrative proliferation of ducts and cords with eccrine or apocrine features extending into the deep dermis and the subcutis. Perineural invasion is also commonly found.³
- Benign syringomas, by contrast, are similar but superficial, less infiltrative, and they do not display perineural invasion.⁴

Case Presentation

- A 20-year-old male presented with two poorly demarcated erythematous nodules on his right infraorbital region and left zygomatic arch, 2.5 cm and 3.2 cm, respectively (Figure 1).
- The nodules were stable for a combined 6 years.
- He had no other similar lesions. Due to their visibility, patient worried about expansion and new lesion formation.
- No previous personal or family medical history of such lesions was reported.



Fig. 1 Showing nodular erythematous swelling on the right infraorbital region and left zygomatic arch

Physical examination revealed two slightly indurated, poorly demarcated, salmon-colored nodules on the right infraorbital region and left zygomatic arch, measuring 2.5 cm and 3.2 cm, respectively (Figure 1).

Histologic Findings

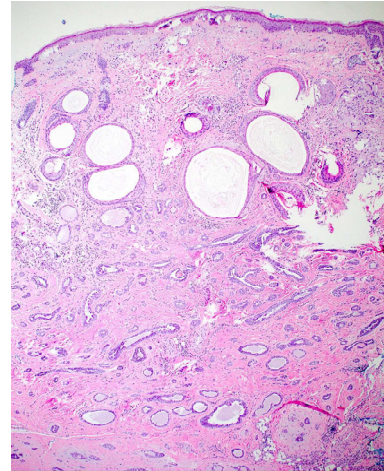


Fig. 2 Histopathology from the left zygomatic arch incisional biopsy showing the keratocysts in the upper dermis and the cords and ducts infiltrating into the deep dermis and subcutis (H&E 4X).

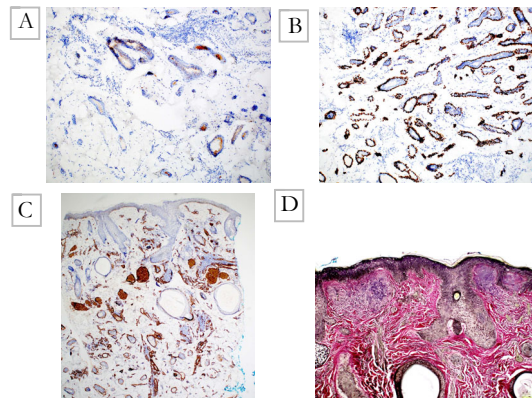


Fig. 3 Immunohistochemical staining of left zygomatic arch incisional biopsy

- A. **EMA:** Marks the luminal cytoplasm and cell membrane of some of the ducts, consistent with apocrine differentiation corresponding to the luminal apocrine budding seen on H&E-stained tissue sections (10X)
- B. **P63:** Stains the inner basal myoepithelial cell layer of the infiltrative ducts (10X)
- C. **SMA:** Stains the inner myoepithelial cell layer of the infiltrative ducts and pillar muscle (4X)
- D. **Verhoeff-van Gieson:** Marks the ball of elastic fibers in the upper papillary dermis (10X)

Clinical Course

- Excisional biopsies of the nodules were taken for a comprehensive work up.
- On H&E staining, the nodules mimicked features of microcystic adnexal carcinoma (MAC). However, the totality of the pathologic features, including staining patterns of p63 and SMA combined with a lack of perineural invasion, suggested a benign disease process.^{5,6}
- Patient was informed of findings and the clinical suspicion that clear margins were unnecessary. Patient was advised to continue monitoring.
- Approximately one year later, no progression has been reported.

Discussion

- Histopathologically, MAC is characterized by small keratocysts in the upper dermis with a deep infiltrative proliferation of ducts and cords with eccrine or apocrine features extending into the deep dermis and subcutis. Perineural invasion is commonly found.^{3,5,6}
- Histopathologic features from excisional biopsies mimicked microcystic adnexal carcinoma (MAC), including the keratin microcysts with deep dermal and subcutaneous infiltration of the keratin-positive cords and ducts of approximately two cell thickness (Figures 2). No perineural invasion was found.
- EMA immunohistochemical stains marked the cytoplasm/membranes of some of the outer duct-like structures, but the CEA was negative, supporting apocrine and not eccrine differentiation (Figure 3A). Unlike MAC, the ductal proliferations had basaloid myoepithelial cells, which were positive on p63 and SMA (Figures 3B & 3C), as well as myoid to collagenous stromal proliferations around some ducts. In addition, there were ball-like aggregates of elastic fibers in the papillary dermis (Figure 3D).
- Thus, the **clinical and histopathologic features supported a benign deep syringoid ductal proliferation (DSDP)**. Similar cases have been previously reported and demonstrated germline pathogenic variants in MYH9.⁷⁻⁸
- Benign syringomas are similar but more superficial, less infiltrative, and lack perineural invasion.⁴
- Such histopathology has been reported with Nicolau-Balus, Rombo, and eccrine pilar hamartomas syndromes.⁷⁻⁸
- Recognizing DSDP is crucial for accurate diagnosis and patient outcomes, as MAC may require a disfiguring operation.

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