Plaquelike Syringoma Mimicking Microcystic Adnexal Carcinoma: A Potential Histologic Pitfall

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PRACTICE **POINTS**

- Dermatologists should familiarize themselves with the plaquelike subtype of syringoma, which can histologically mimic the superficial portion of microcystic adnexal carcinoma (MAC).
- Careful recognition of plaquelike syringoma in the Mohs micrographic surgery setting may prevent unnecessary surgical morbidity.
- Further diagnostic investigation is warranted for superficial biopsies suggestive of MAC or when other characteristic features are lacking.

To the Editor:

Plaquelike or plaque-type syringoma is a lesser-known variant of syringoma that can appear histologically indistinguishable from the superficial portion of microcystic adnexal carcinoma (MAC). The plaquelike variant of syringoma holds a benign clinical course, and no treatment is necessary. Microcystic adnexal carcinoma is distinguished from plaquelike syringoma by an aggressive growth pattern with a high risk for local invasion and recurrence if inadequately treated. Thus, treatment with Mohs micrographic surgery (MMS) has been recommended as the mainstay for MAC. If superficial biopsy specimens reveal suspicion for MAC and patients are referred for MMS, careful consideration should be made to differentiate MAC and plaquelike syringoma early to prevent unnecessary morbidity.

A 78-year-old woman was referred for MMS for a left forehead lesion that was diagnosed via shave biopsy as a desmoplastic and cystic adnexal neoplasm with suspicion for desmoplastic trichoepithelioma or MAC (Figure 1). Upon presentation for MMS, a well-healed, 1.0×0.9-cm scar at the biopsy site on the left forehead was observed (Figure 2A). One stage was obtained by standard MMS technique and sent for intraoperative processing (Figure 2B). Frozen section examination of the first stage demonstrated peripheral margin involvement with syringomatous change confined to the superficial and mid dermis (Figure 3). Before proceeding further, these findings were reviewed with an in-house dermatopathologist, and it was determined that no infiltrative tumor, perineural involvement, or other features to indicate malignancy were noted. A decision was made to refrain from obtaining any additional layers and to send excised Burow triangles for permanent section analysis. A primary linear closure was performed without complication, and the patient was discharged from the ambulatory surgery suite. Histopathologic examination of the Burow triangles later confirmed findings consistent with plaquelike syringoma with no evidence of malignancy (Figure 4).

Syringomas present as small flesh-colored papules in the periorbital areas. These benign neoplasms previously have been classified into 4 major clinical variants: localized, generalized, Down syndrome associated, and familial.[1] The

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The authors report no conflict of interest.

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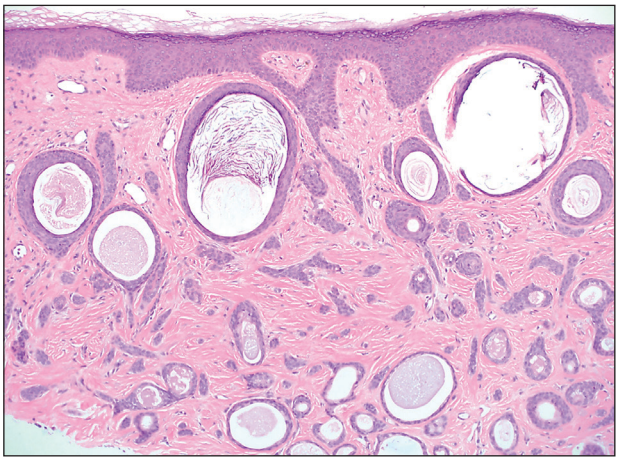

FIGURE 1. Histopathology of an initial shave biopsy permanent section of a left forehead lesion showed a transected ductal proliferation (H&E, original magnification ×100).

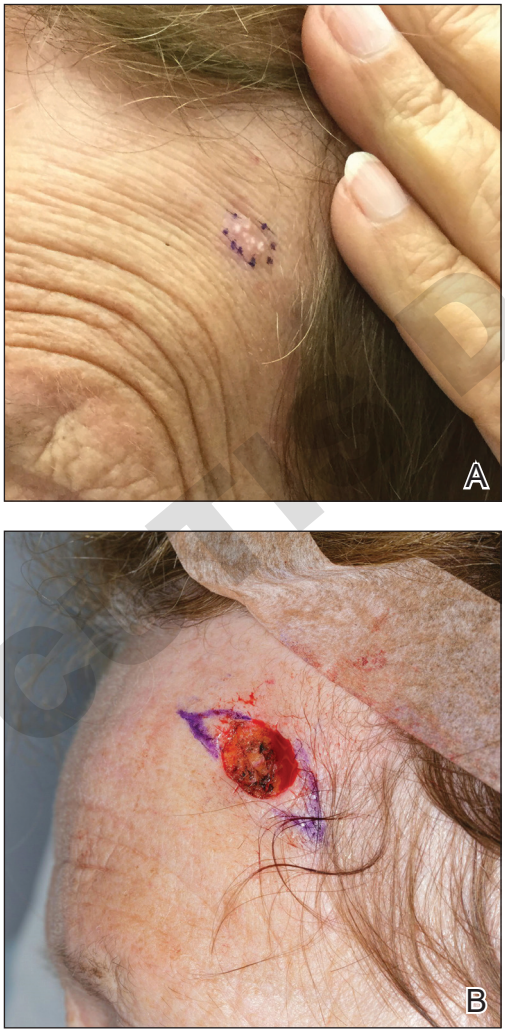

FIGURE 2. A, Prior to biopsy, examination revealed an ill-defined, flesh-colored to white, smooth plaque on the left forehead. B, After 1 stage of Mohs micrographic surgery with Burow triangles drawn.

lesser-known plaquelike variant of syringoma was first described by Kikuchi et al[2] in 1979. Aside from our report, a PubMed search of articles indexed for MEDLINE using the terms *plaquelike* or *plaque-type syringoma* yielded 16 cases in the literature.[2-14] Of these, 6 were referred to or encountered in the MMS setting.[8,9,11,12,14] Plaquelike syringoma can be solitary or multiple in presentation.[6] It most commonly involves the head and neck but also can present on the trunk, arms, legs, and groin areas. The clinical size of plaquelike syringoma is variable, with the largest reported cases extending several centimeters in diameter.[2,6] Similar to reported associations with conventional syringoma, the plaquelike subtype of syringoma has been reported in association with Down syndrome.[13]

Histopathologically, plaquelike syringoma shares features with MAC as well as desmoplastic trichoepithelioma

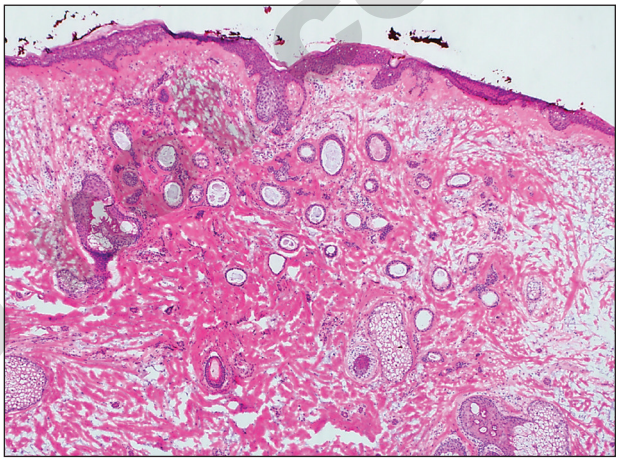

FIGURE 3. Histopathology of an intraoperative frozen section at the first stage of Mohs micrographic surgery showed peripheral margins with syringomatous change within the superficial and mid dermis (H&E, original magnification ×40).

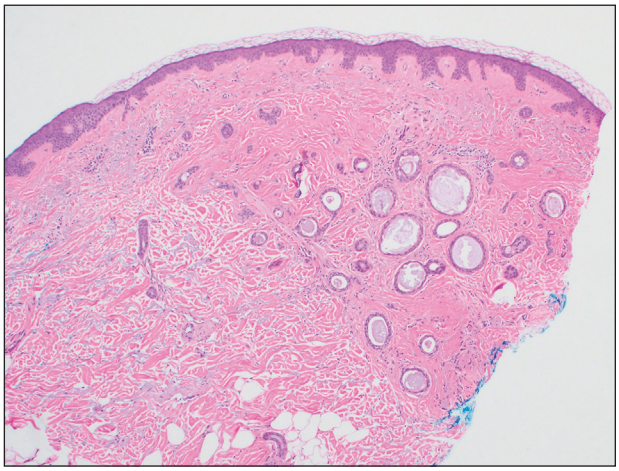

FIGURE 4. Histopathology of a postoperative permanent section from an excised Burow triangle showed only findings of plaquelike syringoma and no infiltrative component (H&E, original magnification ×40).

and desmoplastic basal cell carcinoma. Plaquelike syringoma demonstrates broad proliferations of small tubules morphologically reminiscent of tadpoles confined within the dermis. Ducts typically are lined with 2 or 3 layers of small cuboidal cells. Microcystic adnexal carcinoma typically features asymmetric ductal structures lined with single cells extending from the dermis into the subcutis and even underlying muscle, cartilage, or bone.[8] There are no reliable immunohistochemical stains to differentiate between these 2 entities; thus, the primary distinction lies in the depth of involvement. Desmoplastic trichoepithelioma is composed of narrow cords and nests of basaloid cells of follicular origin commonly admixed with small cornifying cysts appearing in the dermis.[8] Colonizing Merkel cells positive for cytokeratin 20 often are present in desmoplastic trichoepithelioma and not in syringoma or MAC.[15] Desmoplastic basal cell carcinoma demonstrates narrow strands of basaloid cells of follicular origin appearing in the dermis. Desmoplastic trichoepithelioma and desmoplastic basal cell carcinoma are each fundamentally differentiated from plaquelike syringoma in that proliferations of cords and nests are not of eccrine or apocrine origin.

Several cases of plaquelike syringoma have been challenging to distinguish from MAC in performing MMS.[8,9,11] Underlying extension of this syringoma variant can be far-reaching, extending to several centimeters in size and involving multiple cosmetic subunits.[6,11,14] Inadvertent overtreatment with multiple MMS stages can be avoided with careful recognition of the differentiating histopathologic features. Syringomatous lesions commonly are encountered in MMS and may even be present at the edge of other tumor types. Plaquelike syringoma has been reported as a coexistent entity with nodular basal cell carcinoma.[12] Boos et al[16] similarly reported the presence of deceptive ductal proliferations along the immediate peripheral margin of MAC, which prompted multiple re-excisions. Pursuit of permanent section analysis in these cases revealed the appearance of small syringomas, and a diagnosis of benign subclinical syringomatous proliferations was made, averting further intervention.[16]

Our case sheds light on the threat of commission bias in dermatologic surgery, which is the tendency for action rather than inaction.[17] In this context, it is important to avoid the perspective that harm to the patient can only be prevented by active intervention. Cognitive bias has been increasingly recognized as a source of medical error, and methods to mitigate bias in medical practice have been well described.[17] Microcystic adnexal carcinoma and plaquelike syringoma can be hard to differentiate especially initially, as demonstrated in our case, which particularly illustrates the importance of slowing down a surgical case at the appropriate time, considering and revisiting alternative diagnoses, implementing checklists, and seeking histopathologic collaboration with colleagues when necessary. Our attempted implementation of these principles, especially early collaboration with colleagues, led to intraoperative recognition of plaquelike syringoma within the first stage of MMS.

We seek to raise the index of suspicion for plaquelike syringoma among dermatologists and dermatologic surgeons, especially when syringomatous structures are limited to the superficial dermis. We encourage familiarity with the plaquelike syringoma entity as well as careful consideration of further investigation via scouting biopsies or permanent section analysis when other characteristic features of MAC are unclear or lacking. Adequate sampling as well as collaboration with a dermatopathologist in cases of suspected syringoma can help to reduce the susceptibility to commission bias and prevent histopathologic pitfalls and unwarranted surgical morbidity.

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