Sclerosing sweat duct carcinoma of the lower extremity treated with Mohs micrographic surgery

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INTRODUCTION

Sclerosing sweat duct carcinoma (SSDC) is a rare slow-growing malignant tumor of the sweat glands that usually occurs in the head and neck region.[1,2] SSDC is often misdiagnosed clinically, which can result in tumor mismanagement and recurrence.[3] Recent literature has shifted in favor of Mohs micrographic surgery (MMS) as the optimal approach for this type of tumor because of its tissue-sparing effects and high cure rates.[4,5] We present an unusual case of SSDC of the lower extremity that was successfully treated with MMS.

Abbreviations used:

MMS: Mohs micrographic surgery
SSDC: sclerosing sweat duct carcinoma

CASE REPORT

A 59-year-old healthy white man presented with a 3-year history of a slowly enlarging growth over the right Achilles tendon. The patient thought that this lesion was a benign callus on his skin and did not seek immediate treatment. He was otherwise healthy, reported no trauma to the area, and complained of no constitutional symptoms.

On examination, the patient exhibited a well-circumscribed 2.5- × 2.0-cm firm yellow pink tumor located on the right posterior calf overlying the Achilles tendon (Fig 1). No lymphadenopathy was palpable in the right popliteal or inguinal areas.

An incisional skin biopsy found cords and nests of basaloid and atypical cells with ductal structures infiltrating a sclerotic dermis (Fig 2). Perineural invasion was present (Fig 3). Immunohistochemical stains were positive for AE1/AE3, p63, and epithelial membrane antigen and negative for carcinoembryonic antigen. A consensus diagnosis of SSDC was made.

After the biopsy, magnetic resonance imaging was performed to assess the degree of tumor invasion. No Achilles tendon or bony structure involvement was noted. This finding excluded the need for orthopedic surgery and made MMS a viable option for this patient.

Tumor removal was accomplished with 1 stage of MMS using 5-mm surgical margins. The final tumor defect measured 3.5 × 3.8 cm. Because of the high degree of skin tension, a trilobed transposition flap was used to close the defect (Fig 4). An Unna wrap was subsequently applied to assist with healing.

After MMS, the patient was referred to a surgical oncologist. Findings from a comprehensive workup were normal, and the patient was able to ambulate in short order without difficulty. At his 10-month follow-up appointment, the patient had no clinical evidence of tumor recurrence. He was subsequently lost to follow-up despite multiple attempts to contact him.

DISCUSSION

SSDC is a rare malignant tumor first described by Goldstein in 1982.[6] It is thought to be synonymous

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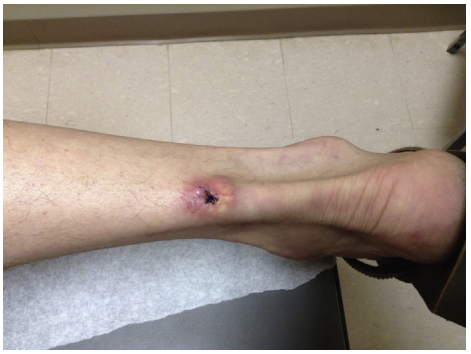

Fig 1. Sclerosing sweat duct carcinoma. A 2.5-cm pale yellow tumor over the superior aspect of the right Achilles tendon.

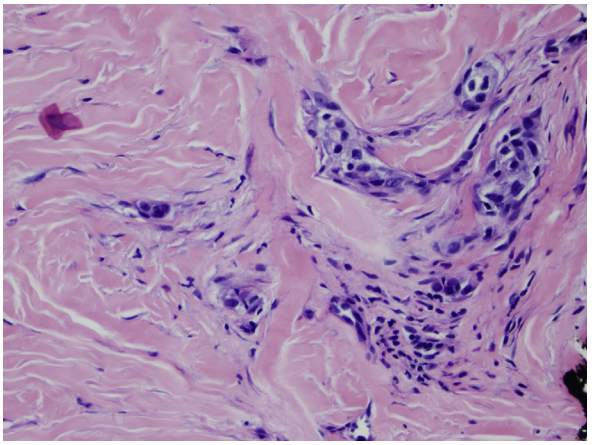

Fig 2. The carcinoma infiltrates a sclerotic dermis in cords and solid aggregates with ductal structures present (56 × 42 mm; 96 × 96 DPI). (Hematoxylin-eosin stain; original magnification: ×200.)

with microcystic adnexal carcinoma[6] and is a type of sweat gland neuroendocrine tumor.

SSDC usually occurs in whites in the fourth to sixth decade of life and has an equal sex distribution.[5,7] Clinically, it presents as an indurated nonhealing tumor in which periods of rapid growth punctuate durations of slow growth.[7] Up to 88% of cases occur in the head and neck area,[8] especially on the left side of the face.[9] Although not formally studied, some authors feel that this may be caused by a higher incidence of ultraviolet radiation exposure in this area.[2,3] SSDC has also been observed, less commonly, in the upper extremity, axilla, trunk, and lower extremity.

Histologically, SSDC comprises cords and nests of basaloid cells in desmoplastic stroma,[1,7] which form ductal structures of 1 to 2 cell layers thick. Keratin-filled cysts have also been observed.[7] Immunohistochemical stains with carcinoembryonic antigen, epithelial membrane antigen, pan-cytokeratin, and p63 help confirm tumor identification.[10,11] Perineural invasion

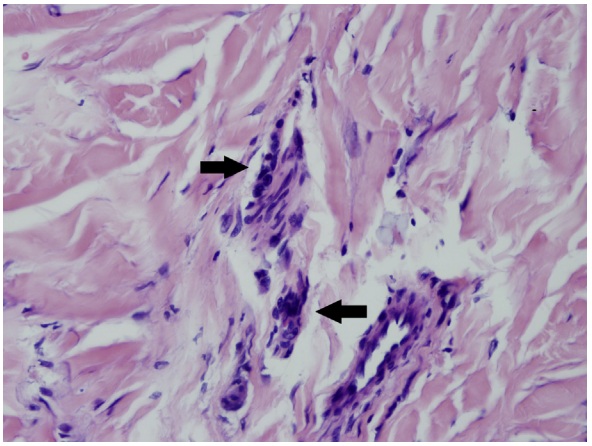

Fig 3. Tumor shows perineural invasion. (Hematoxylin-eosin stain; original magnification: ×400.)

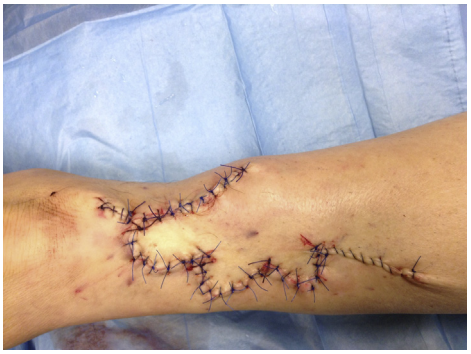

Fig 4. Closing of the defect using a trilobed transposition flap.

is common[4] with up to 80% of lesions having this finding.[1]

In the differential diagnosis, one must consider tumors such as basal cell carcinoma, squamous cell carcinoma, desmoplastic trichoepithelioma, metastatic carcinoma,[7] and syringoma.[4] SSDC has a misdiagnosis rate of up to 27%.[3] Deeper tissue samplings are helpful to minimize false-negative findings.[2,3]

With respect to treatment, there are no randomized prospective studies for this type of tumor, but because there is a slight risk of metastases, the goal should be to completely remove this malignancy from the onset. Margins for excision vary from a few millimeters with MMS to 3 to 5 cm with wide local excision. No clear set standard for margins has been reported in the literature. Local recurrence rates with wide local excision have been reported as high as 40% to 60%.[12] Pathologic "skip areas"[13,14] perineural invasion,[12,15] and the misinterpretation of atypical epithelioid structures as normal[16] are thought to contribute to this observance. Most SSDC recurrences occur within 2 years after treatment,[4,16]

whereas others happen decades later.[17] Careful surveillance of tumor sites postoperatively is recommended. Metastasis from SSDC is rare[16,18] with one death reported in an immunocompromised host.[12]

MMS is the preferred treatment for SSDC,[3,12] as this procedure is found to be a safe, cost effective, and tissue sparing for tumors that grow by local invasion.[18] Literature on the use of adjuvant chemotherapy is limited.[19] Postoperative radiation is not recommended to treat this tumor.[1,6]

With respect to sentinel lymph node biopsy, no formal guidelines exist for tumor staging. Case reports have identified a role for sentinel lymph node biopsy in staging SSDC[15]; however, further long-term randomized studies are still needed to determine if this should be a standard approach to care.

CONCLUSION

SSDC is a challenging tumor to diagnose and treat. It is important to be aware that this malignancy can develop over the lower extremity and to promptly perform a biopsy on any calloused growth or thickening of the skin that does not respond to conventional treatment.

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