Subungual Syringoid Eccrine Carcinoma of the Great Toe Nail Complex

A Case Report

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Syringoid eccrine carcinoma is a very rare skin cancer. We present a case of a 22-year-old woman with a presentation of syringoid eccrine carcinoma in the subungual region of the hallux. This clinical case demonstrates our work-up that led to a proper diagnosis and management of this pathology. We discuss our surgical procedure of choice and the outcome. This report adds valuable information to a limited database of knowledge available on the diagnosis and management of syringoid eccrine carcinomas. (J Am Podiatr Med Assoc 104(5): 504-507, 2014)

A very rare type of skin cancer, primary eccrine carcinoma, accounts for less than .01% of all skin cancers. Primary carcinomas of the adnexal structures of the skin are very rare. Syringoid eccrine carcinoma (SEC) is exceedingly rare and is an uncommonly diagnosed malignancy.[1] These carcinomas are typically locally invasive and destructive, often recur, and up to 60% metastasize, resulting in mortality.[2] Ohnishi et al[3] report that there is no confirmed origin or differentiation for SEC, and the tumor mostly comprises small cords and nests from the reticular dermis to the subcutaneous tissue. In some cases SECs are described as numerous dilated and branching tubular structures of the fibrocollagenous matrix.[4] Our review of literature revealed no case reports of syringoid eccrine carcinoma of the nail complex. In addition, to our knowledge, there were no cases of syringoid eccrine carcinomas reported in the lower extremity. However, many cases have been reported of SEC elsewhere with the only successful treatment being surgical excision of the tumor.[1,4] We report an unusual presentation of subungual syringoid eccrine carcinoma involving the great toe of a 22-year-old female patient.

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Case Report

A 22-year-old woman was evaluated at the Illinois Center for Foot and Ankle Surgery for the first time in February of 2006. She related pain in her great toe underneath the nail. Her personal medical history was unremarkable, but her family history was significant for a second-degree relative (an uncle) with a history of malignant melanoma. The patient was in good physical shape and without any constitutional symptoms.

Twelve months prior to our evaluation, she was treated for suspected onychomycosis of the painful nail with oral antifungals without success. During this period, she reported increased pain and erythema to the periungal and subungual region of this toe. At her first examination at our institution, a soft-tissue mass was noted in the subungual area of the left great toe. The mass extended dorsally, leading to lifting of the nail and encompassing the entire nail bed. The mass appeared to be localized with no associated lymphadenopathy (Fig. 1).

Routine laboratory tests (sedimentation rate, complete blood cell count with differentiation, and liver profile) were all normal or negative. Plain radiographs were negative (Fig. 2). Magnetic resonance images of the left foot from January, 2006 was read as negative for pathology regarding the area of the soft-tissue mass; degenerative joint disease of the first metatarsophalangeal joint was noted.

Because of the patient's family history, significant

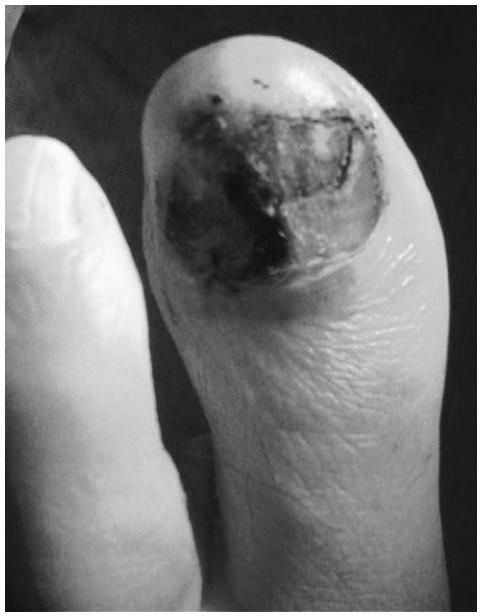

Figure 1. Photograph showing preoperative clinical presentation.

pain, and atypical appearance of this subungual soft-tissue mass, an excisional biopsy was performed with a bone biopsy of the adjacent phalanx in March, 2006. The biopsy of the subungual mass from the left hallux was sent in formalin, and consisted of a 1.8 × 1.2 × 0.5-cm tan-pink, rubbery soft-tissue fragment with surrounding tan skin, and a detached 1.8 × 1.4 × 0.3-cm toenail. Representative sections were submitted for histologic sections. Separately received in formalin designated as bone biopsies of the left hallux, were multiple fragments of tan-gray firm bone, measuring 1.1 × 0.9 × 0.3 cm in aggregate. They were submitted entirely after decalcification.

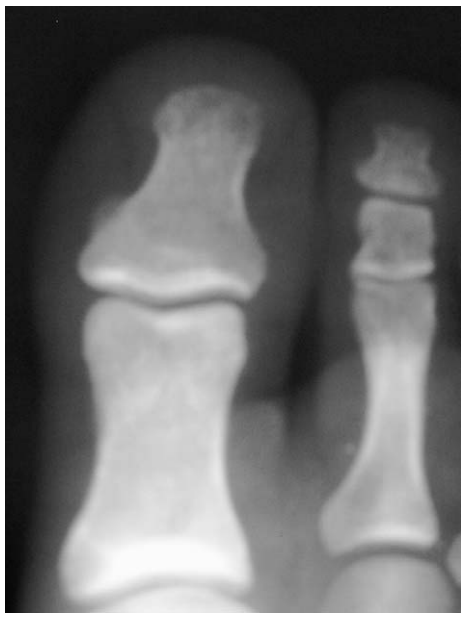

Figure 2. A lateral oblique radiographic view of preoperative evaluation.

Microscopic evaluation revealed a poorly circumscribed proliferation of round to elongated "eccrine-type" ducts in a background of a fibrotic, desmoplastic-appearing stroma. The ducts were lined by one to two layers of cuboidal cells with small, round nuclei and ill-defined pale cytoplasm. Small ducts with tail-like extensions were characteristic (Fig. 3). The cells were positive by immunohistochemistry for carcinoembryogenic antigen (CEA) (Fig. 4), and positive for cytokeratin AE1/AE3. They were negative for epithelial membrane antigen (EMA) and S-100. Scattered clusters of S-100–positive nerve fibers were identified in the background, along with a proliferation of glomus-type cells. The lesion was seen extending to the margins of the specimen. The separately sent fragments of bone revealed replacement of the bone marrow with desmoplastic appearing stroma containing rare epithelial-like structures, suggestive of bone involvement by the tumor (Fig. 5). The final diagnosis was well-differentiated syringoid carcinoma, arising in the background of an eccrine/stromal hamartoma, with increased glomus bodies, and neural hamartomas with desmoplastic response.

A repeat magnetic resonance image after the excisional biopsy revealed post-surgical changes

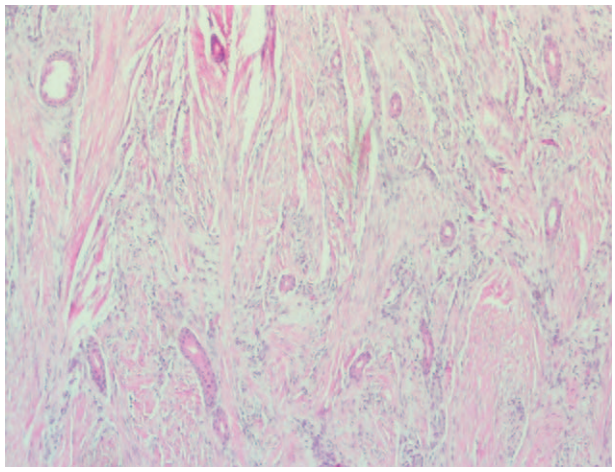

Figure 3. Photomicrograph showing the tumor composed of an admixture of small epithelial nests, thin cords and strands, cystic and ductular structures, proliferating in an eosinophilic fibrotic stroma (H&E, x10).

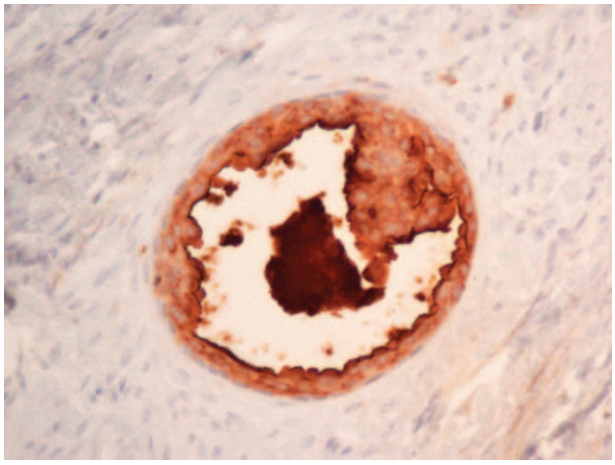

Figure 4. Photomicrograph showing cells positive for carcinoembryogenic antigen (CEA) (H&E, x40).

with no evidence of bony invasion. Based on the diagnosis of syringoid eccrine carcinoma, a wide excision was planned. At this point, the lesion appeared hypergranular with areas of necrosis and atypia, measuring 2.5 × 2.1 cm. The lesion was excised with a 0.5-cm clear margin (Fig. 6). The distal tuft of the distal phalanx was resected and an additional clearance fragment was obtained. Bioengineered skin substitute was applied afterward. Histopathology revealed clear margins of the soft tissue and bone.

Discussion

Although SEC is a rare tumor in the lower extremity, clinicians, in general, should keep malig-

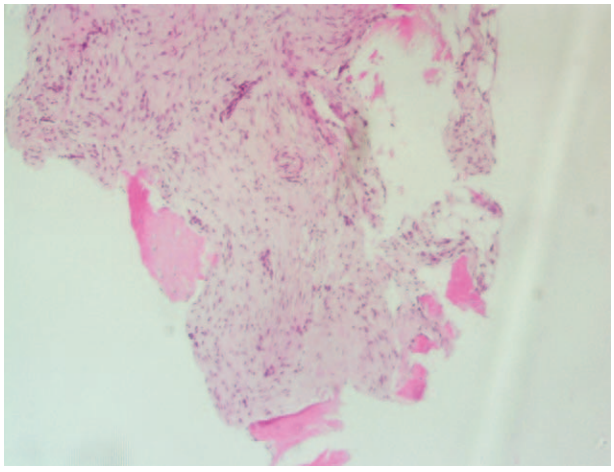

Figure 5. Photomicrograph showing thin cords of epithelial-like structures and fibrotic stroma surrounding bony trabeculae (H&E, x4).

Figure 6. Postoperative photo of surgical site.

nancy high on their list of differential diagnoses. It is especially crucial when they are dealing with a recalcitrant lesion that did not respond or appeared worse after initial treatment was rendered. In this particular case, a high index of suspicion for malignancy was the main reason of aggressive work-up and proper diagnosis. Although this type of tumor recurs frequently, we propose a treatment plan to prevent the recurrence. Currently, the tumor in our patient has not recurred since the date of surgery, and with no other treatment guidelines available for SEC in the lower extremity in the subungual area, we report successful surgical

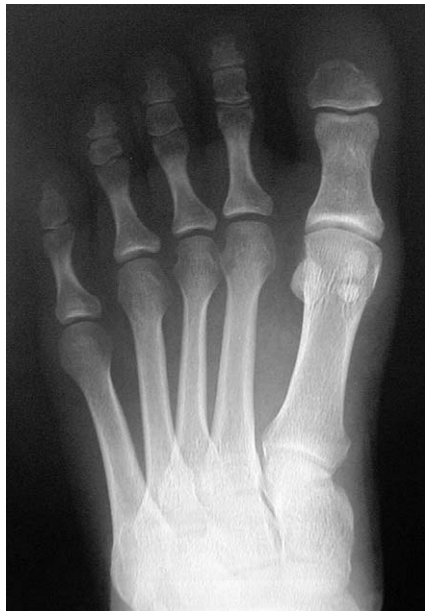

Figure 7. A dorsoplantar radiographic view of postoperative evaluation.

excision of the tumor with a 0.5-cm clear margin and excision of adjacent bone due to presence of reactive changes with good long-term outcome (Fig. 7). With the significant metastatic rate and histopathologic findings of SEC given,[2] it is essential to treat eccrine carcinomas suitably. Our case demonstrates the technique of diagnosing and treating SEC in the lower extremity, which has never before been reported.

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Conflict of Interest: None reported.

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