Sclerosing Syringoid Carcinoma of the Finger

Chuieng-Yi Lu², MD; Shih-Ming Jung¹, MD; Yu-Te Lin², MD

We present a case of sclerosing syringoid carcinoma of the finger, a type of eccrine sweat duct malignancy that is rare and mimics the appearance of a benign lesion, but is locally aggressive and has a tendency to recur. It has seldom been reported in the literature. It has a predilection for the head and neck region but is rare in the extremities. We believe this is the second reported case of sclerosing syringoid carcinoma in the finger, after the diagnosis was determined from a histological and immunohistochemical study. The disease presentation and the difficulties encountered during establishment of the diagnosis and planning of the treatment are discussed. (Chang Gung Med J 2011;34:115-21)

Key words: sclerosing syringoid carcinoma, syringoid eccrine carcinoma, finger, digit

Sclerosing syringoid carcinoma is a rare type of eccrine sweat duct malignancy. It was first introduced by Lipper and Peiper in 1979 as a type of sweat gland carcinoma with syringomatous features, and later defined in larger case series by Mehregan et al in 1982 and Cooper et al in 1985.(1-3) It tends to invade deep tissues such as the perivascular or perineural tissue, which is one of the main differences that separates it from the benign syringoma. The malignant form is known to easily recur if not adequately excised. Because of its predilection for craniofacial regions, it has been met with growing awareness for malignancy when encountered in clinical situations.(2-4) However, it has rarely been reported in the extremities and can be easily disguised as a firm painless mass with a typical benign presentation. Here we present a rare case which may be the second case of sclerosing syringoid carcinoma reported on a finger.

CASE REPORT

A 47 year-old woman presented to the outpatient clinic with a protruding mass distal to the 2nd metacarpo-phalangeal joint on the ulnar side of the left index finger that had been present for over 5 years (Fig. 1). Initially the mass was only palpable subcutaneously, and was firm, moveable, and painless upon palpation. The mass gradually grew larger, and then ulcerated through the overlying skin, with an indurated irregular contour and overlying telangiectasia. She denied numbness or cold intolerance in the index finger and had no other similar lesions on her body. She denied a past history of chronic diseases or recent traumatic accidents. She denied any family history of soft tissue malignancies but noted that her father had suffered from a brain tumor and her mother had died recently of lung adenocarcinoma. The initial differential diagnosis included common soft tissue tumors of the hand such as ganglions linked to the joint capsule or giant cell tumors of the tendon sheath. Upon exploration under local anesthesia, the mass was lobulated and well capsulated overlying the ulnar digital neurovascular bundles, and it was easily separated from the surrounding soft tissue (Fig. 2). Under the impression of a benign soft tissue tumor, local excision of the mass with primary suture of the wound was performed and the specimen was...
sent for further pathological evaluation.

The final pathology report of the mass revealed syringoid carcinoma of the left index finger, a type of eccrine sweat duct malignancy that has rarely been reported on the extremities. The mass submitted was 1.8 x 1.5 x 1.1 cm and microscopic examination revealed infiltrative nests of malignant glandular cells immersed within a desmoplastic stroma (Fig. 3). The surgical margin of the specimen was involved. The tumors cells were stained with immunohistochemical stains D2-40 and P63 and the final pathological diagnosis was sclerosing syringoid carcinoma.

Wide excision of the residual left index wound was then performed and the plan was to excise all tissues involved, including the tendon and neurovascular bundles of the left index finger if required. The specimen, which was clinically the peripheral margin of the previous wound, was sent for intra-operative frozen section and showed a negative margin. A subsequent free arterialized venous flap was used for reconstruction of the wound defect on the left 2nd index finger (Fig. 4). In addition, sentinel lymph node mapping was performed prior to the operation and a lymph node was dissected at the posterior margin of the pectoralis major. After the operation, the left index flap wound healed without any major complications. The final histological result did not find any residual tumors in the specimen with at least 5 mm of peripheral clearance and the dissected lymph node was negative for malignancy. At 6 months postoperatively, there was no palpable mass or any signs of recurrence during follow-up visits (Fig. 5).
DISCUSSION

In this case report, we used the term sclerosing syringoid carcinoma to emphasize the findings of infiltrative nests of basaloid cells, derived from the ductal cells of the intradermal eccrine sweat glands, immersed within a desmoplastic stroma. A review of the current literature showed many synonyms including sclerosing sweat duct carcinoma, microcystic adenexal carcinoma, syringoid eccrine carcinoma, malignant syringoma and malignant eccrine epithelioma. In some reports, the authors view the term sclerosing sweat duct carcinoma as synonymous with microcystic adnexal carcinoma, which is a type of multiphasic differentiated tumor presenting with the characteristic findings of superficial keratinous cysts and a deeper part with nests of glandular cells in dense sclerotic stroma. Cooper et al divided 20 patients with sclerosing sweat duct carcinoma into two groups, one with disease more comparable to microcystic adnexal carcinoma introduced by Goldstein et al in 1982, and the other with disease more similar to syringoid eccrine carcinoma introduced by Lipper et al in 1979 and Mehregan et al in 1983. However, the classification of microcystic adnexal carcinoma, which was originally perceived as having an eccrine origin, has been complicated by reported findings of apocrine and pilosebaceous differentiation, which may suggest that this tumor has a mixed adnexal lineage. Cooper et al divided 20 patients with sclerosing sweat duct carcinoma into two groups, one with disease more comparable to microcystic adnexal carcinoma introduced by Goldstein et al in 1982, and the other with disease more similar to syringoid eccrine carcinoma introduced by Lipper et al in 1979 and Mehregan et al in 1983. We found no superficial keratinous cysts in our patient. To confirm the eccrine glandular origin, certain markers can be used, such as the enzymes PAS, succine dehydrogenase and leucine aminopeptidase, which react strongly with cells of eccrine glandular origin but mildly with apocrine cells. Also, immunohistochemical stains such as CEA and S-100 may aid in the diagnosis. We prefer to add the term ‘syringoid’ to our diagnosis, emphasizing the involvement of eccrine ductal cells and similarities to the benign syringoma. This tumor may still be viewed as a subtype of microcystic adnexal carcinoma without the superficial keratinous cyst component, but for purposes of emphasizing the malignant transformation of eccrine ductal cells in desmoplastic stroma, we will refer to the term sclerosing syringoid carcinoma in the following discussion.

Sclerosing syringoid carcinoma is not only one of the rarest skin appendage tumors, but is also seldom reported among eccrine sweat gland malignancies. It has been found more often in the craniofacial regions, such as the lips, eyelids, nose, nasolabial fold, chin, ear and scalp. It is much less common
in regions outside of the head and neck, with cases reported in the axilla, the trunk, and rarely the extremities.\(^{2,3,6,11,12}\) Most reported cases follow a similar history of a slowly growing papule or an indurated plaque of variable size, covered by an epidermis that can be normal, atrophic or scaly.\(^{2,7}\) The mass is usually firm and solitary and it rarely ulcerates through the skin. However, after close examination under the microscope, the tumor often extends beyond the clinically visible or palpable margins, both in length and depth. This presents a very troublesome situation for clinicians whose initial impression is usually that of a benign cutaneous lesion. In our case, our initial plan was to perform gross dissection with simple excision of the mass, under the suspicion of a benign tumor such as a ganglion or giant cell tumor. However, most cutaneous sweat duct malignancies can only be remotely removed by Moh's micrographic surgery or by wide excision combined with frozen section confirmation, and regional lymphadenectomy is recommended if lymph node metastasis is suspected.\(^{10,15,16}\) The treatment result is often difficult to predict because first, the tumor is poorly circumscribed and it may invade the deep dermis right down to the underlying muscle or perivascular tissue, leading to significant soft tissue loss if a true complete wide excision is planned.\(^{2,3,6,11}\) It is furthermore characterized by dispersed and discontinuous growth, which may lead to unreliability of the surgical margins for the layer by layer approach of current treatment strategies.\(^{2,6}\) It is highly recurrent, ranging from 30% to 47%, and may recur at the original site even after a long disease-free interval of 30 years.\(^{2,3,17}\) The ideal treatment therefore requires an extensive excision with surgical margins of adequate length and depth. Sinha et al suggested a surgical margin of at least 2 cm for syringoid eccrine carcinoma, but as their report and others indicated, even after generous clinical excision margins of up to 2 cm, the final histological margin was eventually found to be a few millimeters away.\(^{11}\) In concordance with their findings, our final histological surgical margin was only 5 mm, highlighting the limitations of wide excision and perhaps suggesting the need for more precise methods such as Moh's surgery. Secondly, unlike the highly aggressive digital papillary adenocarcinoma or other sweat gland malignancies, this tumor rarely metastasizes to distant organs. However, limited cases of regional lymph node metastasis in syringoid eccrine carcinoma were found in Evan et al and Bogner et al’s case series and only one case of distant metastasis related to syringoid eccrine carcinoma was reported previously.\(^{18,19}\) Lymphatic mapping and sentinel lymph node biopsy have been advocated for other sweat gland neoplasms, but it remains largely unreported for syringoid eccrine carcinomas.\(^{19}\) In our case, there were no physical signs of lymphatic involvement or any suspicion of distant metastasis. However, given the limited information regarding predictors for lymphatic spread and the unpredictable nature of sweat gland neoplasms, we chose to perform sentinel lymph node biopsy to rule out any lymphatic spread.

Our patient presented with an ulcerated mass on the ulnar side of the left index finger that had existed for 5 years. This tumor showed slow growth due to the fact that it did not expand to the adjacent joints or other phalanges. Tumors of the hand can be grouped into their respective origins, epidermal, connective tissue, vascular, and neural, and further divided into benign and malignant presentations. The three most common tumors on the hand are giant cell tumors of the tendon sheath, ganglia (including mucous cysts), and epidermal inclusion cysts.\(^{9}\) The most common hand malignancy is squamous cell carcinoma, followed by other epidermal malignancies but rarely sweat gland malignancies. Other rare soft tissue tumors that present as nodular masses include fibromas, traumatic neuromas and schwannomas, which are all slow growing tumors that are benign in appearance and may become painful. Only 1% to 2% of primary tumors of the hand are malignant, the most common of which is squamous cell carcinoma.\(^{9}\) Given our patient’s presentation of a subcutaneous mass that gradually ulceraeted through the skin, malignancy should still be considered but an origin in the sweat gland was highly unexpected. Eccrine sweat glands are widely distributed almost everywhere on the body, but it is noteworthy that the palms and soles, where the eccrine sweat glands abound, are not the preferred sites of eccrine malignancies.\(^{9}\) Reported eccrine malignancies include porocarcinoma, hidradenocarcinoma, spiradenocarcinoma, cylindrocarcinoma, mucinous carcinoma, adenoid cystic carcinoma, microcystic adnexal carcinoma, and syringoid carcinoma. There is a paucity of eccrine tumors reported on the hand, let alone the
finger, and histochemical, immunochemical and ultrastructural properties are often necessary to make a correct diagnosis. In Mehregan et al’s study, only 2 of 35 cases of primary eccrine adenocarcinoma were found on the hand, with one presenting as eccrine porocarcinoma on the dorsum of the hand, and the other as syringoid eccrine carcinoma on the palm of the hand. (2) Goto et al reported the first case of syringoid eccrine carcinoma on the finger and thus we believe this rare case is the second to be reported in literature (Table 1). (12)

The ideal treatment has not yet been established, with previous reports of Mohs surgery or wide excision of the mass combined with frozen section applied to decrease the high recurrence rate. We chose the latter technique for our patient because of the proximity of the underlying vessels and nerves to the original tumor site, which is even more conspicuous on a digit. Larger dissections or even amputations should be considered if the surgical margins are constantly positive for malignancy. Although the final pathology results were negative for residual tumor, the length of follow up is too short for this outcome to be considered an acceptable result. There is no evidence in the literature that warrants the use of radiotherapy or chemotherapy. (5, 20) Further follow-up of patient progress is needed in this case.

**Conclusion**

In conclusion, we report a rare case of sclerosing syringoid carcinoma on the volar side of the left index finger, believed to be the second syringoid eccrine carcinoma found on the finger. The differential diagnosis of nodular masses of the hand include the more common benign tumors such as ganglions or mucous cysts, giant cell tumors and epidermal inclusion cysts. The rarer malignant tumors should also be considered, such as, in our case, the sweat

Table 1. Sclerosing Syringoid Adenocarcinoma of the Upper Extremities

<table>
<thead>
<tr>
<th>Series</th>
<th>Location</th>
<th>Age / Gender / Race</th>
<th>Duration</th>
<th>Gross appearance size</th>
<th>Microscopic presentation</th>
<th>Enzyme reactions / Immunohistochemical stain</th>
<th>Treatment</th>
<th>Recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mehregan et al</td>
<td>Palm of hand</td>
<td>50 / F / Caucasian</td>
<td>Many years</td>
<td>2 cm Nodular infiltrative plaque</td>
<td>Branching tubular structures lined with basaloid cells and embedded in fibrocollagenous stroma; may involve subcutaneous fat and perivascular or perineural tissue</td>
<td>Central lumen material: PSA positive and Diastase-resistant</td>
<td>–</td>
<td>Local recurrence</td>
</tr>
<tr>
<td></td>
<td>Arm</td>
<td>68 / F / Caucasian</td>
<td>Many years</td>
<td>1.5 cm Nodular infiltrative plaque</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Cooper et al</td>
<td>Axilla</td>
<td>21 / F</td>
<td>1.5 years</td>
<td>3 cm indurated, dermal-subcutaneous nodule</td>
<td>Nests and cords of polygonal or cuboidal cells with tail like extensions; cells aggregate to form lumens that contain eosinophilic material; surrounding dense sclerotic stroma; tumor invasion to perineural or perivascular tissue</td>
<td>Excision</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Axilla</td>
<td>26 / M</td>
<td>1.5 cm dermal-subcutaneous nodule</td>
<td>Excision</td>
<td>Loss to follow up</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Axilla</td>
<td>21 / F</td>
<td>2 years</td>
<td>1.8 cm firm, poorly demarcated brownish tumor on dorsum of left middle finger</td>
<td>Nests and cords of atypical basaloid cells in the fibrous stroma, with eosinophilic material in the lumen. Deep perineural invasion with multiple small cords and nests, surrounded by hyalinized connective tissue</td>
<td>1. Excision (margin 0.5 cm)</td>
<td>Distal recurrence (lymphadenopathy) in forearm and axilla</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Finger</td>
<td>83 / M</td>
<td>10 years</td>
<td>Tumor cells: CEA, epithelial membrane antigen and keratin positive</td>
<td>2. Amputation up to MCP joint</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Abbreviations:** PAS: periodic acid-Schiff; CEA: carcinoembryonic antigen; MCP: metacarpophalangeal.
duct malignancies. Sclerosing syringoid carcinoma is characterized by slow growth and low rates of distant metastasis, but has a tendency to infiltrate deep tissues and recurs unpredictably. Therefore, if found on the finger, an extensive excision with surgical margins of adequate length and depth are required along with a detailed plan outlining possible reconstruction options. Further follow-up is required to validate our choice of treatment.

REFERENCES

手指上的纖維化汗腺管惡性腫瘤

呂烱毅1 容世明1 林有德2

纖維化汗腺管惡性腫瘤是一種罕見卻又易復發的汗腺惡性腫瘤。這種腫瘤在文獻上本來就不常被報導到，而最常發現的位置通常是位於頭頸部，幾乎不會出現在四肢上。我們發現這個案例是文獻上第二個被報導位於手指上的纖維化汗腺管惡性腫瘤，並對其診斷及治療加以探討。(長庚醫誌 2011;34:115-21)

關鍵詞：纖維化汗腺管惡性腫瘤，纖維化汗腺管腫瘤，手指，指端