Myoepithelioma of the Ankle: A Case Report

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Abstract

Myoepitheliomas are rare benign and malignant neoplasms composed of spindled, ovoid, or plasmacytoid cells. These tumors have been reported to occur in salivary glands and cutaneous, subcutaneous, and deep soft tissues of the head and neck, upper and lower extremities, lung, and breast. High-grade myoepitheliomas have a greater propensity to recur compared to their low-grade counterparts. Surgical resection with clear margins is the standard of care for the treatment of both low- and high-grade neoplasms. The aim of this paper is to present an interesting case of myoepithelioma of the ankle presenting as a sarcoma.

Case Description

A 55-year-old female with no significant past medical history presented with left ankle pain after suffering an inversion ankle sprain. She described the initial pain as sharp and very tender to the lateral ankle. Radiographs of the left ankle were negative for any bony pathology and she was treated conservatively for 4 weeks. During this time, she continued to have pain and swelling. She developed a localized soft tissue mass at the retromalleolar area of the peroneal tendons. Physical examination would reveal pain, erythema, and edema over the tendons. Pain was elicited with the peroneal tunnel compression test and active eversion of the foot. There was a soft tissue mass 5 cm proximal and posterior to the peroneal tendons, which was minimally painful with palpation. The mass was firm, non-pulsatile, and non-mobile with direct manipulation. The mass appeared to be deep to the peroneal tendons and possibly within the posterior compartment of the leg (Figure 1).

MRI of the left ankle was reviewed and revealed a split longitudinal tear of the peroneal brevis tendon at the level of the retromalleolar area. In addition, there was a well circumscribed lesion posterior to the peroneal tendons that was diagnosed as a soft tissue sarcoma (Figure 2 & 3). After physical examination and evaluation of the MRI it was determined that urgent resection of the soft tissue mass be performed. Preoperative laboratory tests including a complete blood count with differential and basic metabolic panel were all within normal limits.

A posterior lateral incision was made over the lateral ankle and dissection down to the peroneal tendons was performed. The peroneal brevis tendon had a longitudinal tear and was retubularized. Next, deeper to the peroneal tendons and muscle belly, we identified a firm, white, and somewhat vascular mass that was adhered to the peroneal longus muscle belly and extending into the posterior compartment of the leg (Figure 4).

The mass was carefully dissected from its surrounding attachments with a blunt scissor and was sent for immediate frozen section. Preliminary results of the frozen section revealed atypical cells in a background of hyalinized stroma, which was then referred to permanent. The specimen consisted of a continued well encapsulated dense rubbery mass measuring 7.5 x 2.5 x 1.9 cm in all dimensions (Figure 5).

Histopathology

The lesion was a well circumscribed tumor composed mostly of round cells arranged in cords and pseudoacini set against a manipulation to hyalinized stroma (Figure 6). The cellularity is variable throughout the tumor and there are areas that are only composed of hyalinized stroma. Microcyst containing mucinous fluid are also noted. The neoplastic cells contain clear vacuolated to eosinophilic cytoplasm and round to oval slightly hyperchromatic nuclei. Mitoses are rare. There is no necrosis. Immunostains show patchy but strong positivity for EMA (Figure 7). The specimen was diagnosed as a myoepithelialoma with clean resected margins. The post-operative course was uneventful and she was informed that local recurrence of the tumor is possible.

Figure 1: Preoperative mass

Figure 2: Preoperative MRI

Figure 3: Postoperative MRI

Figure 4: Intraoperative mass

Figure 5: Resected mass

Figure 6: Pseudoacini

Figure 7: EMA positive cells

Figure 8: FISH analysis

Discussion

Myoepithelial cells are usually located in glandular epithelium such as sweat, mammary, lacrimal, skin, and salivary glands. While myoepitheliomas are commonly found in the salivary glands, other locations include the limbs and trunk region. The lesion typically presents in young to middle-aged adults with benign or malignant characteristics. Clinically, the mass is palpable, soft, and non-tender. Myoepitheliomas are a rare tumor, first described as a retroperitoneal tumor in 1995 by Burke and associates. Since this report there have been relatively few myoepitheliomas described in the foot and ankle literature. Immunostaining revealed positive strong positivity for EMA, rare cells positive for S100, and negative for pankeratin, CK5/6, desmin, NFP, p63, and ERG. Myoepitheliomas are more commonly classified as benign tumors. The most reliable criteria for benign character is the absence of cytologic atypia. In this presented case, mitosis was rare and difficult to identify. Recurrence of histologically benign tumors has been associated with a local recurrence of 20% and rarely metastasized.

Gene rearrangement at the EWSR1 (22q12) location is seen in over 90% of soft tissue sarcomas. In this case, FISH analysis did not reveal gene rearrangement at EWSR1 22q12. The clinical significance is unknown, the result does not rule out a malignancy or other genetic changes.

References


