Case Reports and Series

Myoepithelioma of the ankle: A case report

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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. Surgical resection with clear margins is the standard of care for the treatment of both low- and high-grade neoplasms. A 55-year-old female with no significant past medical history presented to an urgent care center with left ankle pain after suffering an inversion ankle sprain. Radiographs of the left ankle were reviewed and were negative for any bony pathology. She continued to have pain, edema, and developed a localized soft tissue mass at the retromalleolar area. MRI showed a soft tissue tumor adjacent to the peroneal tendons. The mass was removed and pathology confirmed it as a myoepithelioma, which is a rare tumor of the lower extremity. Myoepitheliomas are most commonly found in the salivary glands and can have a benign or malignant characteristic. Other common locations include the limbs and trunk region and the lesion typically presents in young to middle aged adults. Recurrence of histologically benign tumors has been associated with a local recurrence of 20% and rarely metastasize. Gene rearrangement at the ESWR1 (22q12) location is seen in 15-40% of soft tissue sarcomas. In this case, FISH analysis did not reveal gene rearrangement at ESWR1 22q12. The clinical significance is unknown and does not rule out a malignancy or other genetic changes.

Case Description

A 55-year-old female with no significant past medical history presented to an urgent care center 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 reviewed and were negative for any bony pathology and she was treated conservatively for 4 weeks. After failing conservative therapy, she continued to have pain, edema, and developed a localized soft tissue mass at the retromalleolar area of the peroneal tendons (Fig. 1).

An MRI was ordered for the left ankle and referral to podiatry for further evaluation was placed. 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 (Figs. 2 and 3).

Physical examination would reveal pain, erythema, and edema over the peroneal tendons. Pain was elicited with peroneal tunnel compression test and active eversion of foot on the ankle. In addition, there was a soft tissue mass about 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. Pulses were intact and there were no neurological deficits or dermatological manifestations to the lower extremity.

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 carefully performed. The peroneal brevis tendon did indeed have a longitudinal split tear and this was repaired using a retubularization technique. Next, deeper to the peroneal tendons and muscle belly we identified a firm, white, and somewhat vascular mass that was somewhat adhered to the peroneal longus muscle belly and extending into the posterior compartment of
the leg (Fig. 4). The mass was carefully dissected from its surrounding attachments with a blunt scissors 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 unoriented well encapsulated dense rubbery mass measuring 7.5 x 2.5 x 1.9 cm in all dimensions (Fig. 5). The incision was copiously irrigated, peroneal retinaculum repaired, and layered closure performed without complications.

Histologically and microscopically the lesion showed a well circumscribed tumor composed mostly of round cells arranged in cords and pseudoacini set against a chondromyxoid to hyalinized stroma. 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 and difficult to be seen. There is no necrosis. Immunostains shows patchy but strong positivity for EMA (Figs. 6 & 7).

The specimen was diagnosed as a myoepithelioma with clean resected margins. The post-operative course was uneventful and she was informed that local recurrence of the tumor is possible. FISH analysis (Fig. 8) shows an abnormal signal in the EWSR1 gene rearrangement, although the EWSR1 gene rearrangement at 22q12 is not identified, which is commonly encountered in soft tissue sarcomas.

**Discussion**

Soft tissue masses should be considered malignant if they have any one of the following characteristics; Increasing size, size >5 cm, location
deep to the deep fascia, and painful. Once a patient is suspected of having a sarcoma, an urgent referral to a diagnostic center for further assessment, imaging, and surgical biopsy should be performed. The standard or preferred imaging technique for soft tissue masses and sarcomas is MRI, but in the advent that MRI is not feasible, a CT or Ultrasound may be considered. Current recommendations suggest that a core biopsy be performed by a sarcoma surgeon or experienced radiologist. Next, after biopsy confirmation of the sarcoma, a chest CT should be evaluated to rule out pulmonary metastasis. It is imperative that the biopsy tract be resected and excised entirely during definitive treatment to prevent any seeding of cancerous cells. Overall, the care of the sarcoma patient should be multidisciplinary in nature with consultations to an orthopedic oncologist or sarcoma specialist.

The standard of care for localized sarcomas is surgical excision with the primary goal to excise the entire tumor with a margin of approximately 1 cm of normal tissue. The situation exists that positive margins return from inadequate resection. The patient should then have additional resection if appropriate margins can be achieved because residual disease proves difficult to contain even with radiotherapy.

In the case presented, collaboration was sought between the radiologist interpreting the MRI and it was communicated to our surgical team that immediate resection of the mass be performed with wide margins preferably. We removed the mass along with approximately 2 cm of surrounding tissue and any abnormal appearing connections or tissue attached to the mass. The mass was sent for immediate frozen section, which returned wide surgically resected margins and deferred for permanent analysis. Analysis of the mass returned a myoepithelial tumor.

Myoepithelial cells are usually located in glandular epithelium such as sweat glands, mammary glands and tissue, lacrimal glands, skin, and salivary glands. Myoepithelial cells are located at or near the base of a gland and when the secretory gland is stimulated, the cells contract to facilitate expulsion of saliva or other glandular components. Myoepitheliomas are most commonly found in the salivary glands and can have a benign or malignant characteristic. Other common locations include the limbs and trunk region and the lesion typically presents in young to middle aged adults. Clinically, the soft tissue 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 metastasize. Gene rearrangement at the ESWR1 (22q12) location is seen in over 90% of soft tissue sarcomas. In this case, FISH analysis did not reveal gene rearrangement at ESWR1 22q12. The clinical significance is unknown, the result does not rule out a malignancy or other genetic changes.
Declaration of competing interest

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Patient Informed Consent Statement

Informed consent was sought from the patient in this study. No institutional review board approval was required or obtained.

References