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CASE REPORT

Subcutaneous Myoepithelioma of Foot Mimicking as Hemangioma: an Unusual Presentation with Review of Literature

Spoorthy Rekha Y.C.[1] · Gale Kathleen[1] · Sunil Kumar Alur[2]

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Introduction

Myoepithelial tumors of soft tissues are rare tumors of uncertain histogenesis included in the "mixed tumor/parachordoma family" of soft tissue tumors (WHO classification). The spectrum of such tumors includes various terminologies based upon anatomic location, such as pleomorphic adenoma in the salivary gland, benign mixed tumor in the skin, and myoepithelial tumor/parachordoma in the soft tissues. Cutaneous myoepitheliomas are extremely rare, benign, dermal tumors located mainly in the subcutaneous and sub-fascial planes of the extremities especially in the muscles of the thigh, groin, calf, upper arm, and forearm. They are seen in patients of all ages with a peak incidence in the second and fourth decades of life [1, 2]. Only a few case reports of myoepithelioma, primarily occurring in the dermis have been described, of which < 10 cases have been reported in the foot and ankle literature. We report a case of subcutaneous myoepithelioma arising over the medial malleolus of the foot of a 70-year-old male.

Case Report

A 70-year-old male presented to surgical OPD with complaints of swelling over the medial malleolus of the left foot with 2 years duration, gradually increasing in size associated with mild dragging pain. On examination, overlying skin appeared unremarkable with a subcutaneous swelling measuring 5 × 4 cm with well-defined borders and partial mobility. Initial evaluation done with the Doppler study revealed a vascular heterogeneous lesion with multiple intralesional hypoechoic foci (phleboliths) suggesting a possibility of hemangioma. Further imaging with an MRI study revealed a well-defined space-occupying lesion in the subcutaneous plane of ankle over the medial malleolus. The lesion was seen in close proximity to the tibialis posterior tendon and showed multiple hypointense areas, indicating intralesional hemorrhage. Differentials considered included space-occupying lesions of the ankle—synovial sarcoma, nodular fasciitis, and peripheral nerve sheath tumor. Excisional biopsy with overlying skin was sent for histopathological examination. Gross examination revealed a gray brown nodular soft tissue measuring 4 × 4 × 3 cm with a fragment of the skin (Fig. 1). External surface appeared capsulated and smooth. Cut surface showed hemorrhage with solid gray tan areas and intervening foci of glistening appearance suggesting cartilage with hard bone–like consistency. Hematoxylin and eosin stained sections revealed a capsulated neoplasm composed of neoplastic cells arranged in sheets, nests, and trabeculae separated by thick fibrous septa. Individual tumor cells with plasmacytoid appearance noted displaying mild nuclear pleomorphism (Fig. 2) with abundant amphophilic cytoplasm and surrounding myxoid change. Areas of mature hyaline cartilage with intervening foci of hemorrhage and ischemic necrosis are seen. The tumor was limited to the capsule with no mitosis and the overlying skin appeared uninvolved by the tumor. Considering its benign nature and its close proximity to the skin, a dermal adnexal tumor, chondroid syringoma was considered as close differential along subcutaneous myoepithelioma, but an underlying possibility of extraskeletal chondrosarcoma could not be ruled out in view of the anatomic location, age of the patient, and the presence of heterologous components like mature cartilage. Chondroid syringoma was excluded based on morphology as the other supporting features like ductal structures, nests of polygonal cells, and keratinous cysts were absent. With the remaining two differentials of subcutaneous

✉ Spoorthy Rekha Y.C.
spoorthyrekha@gmail.com

[1] Department of Histopathology, Elbit Medical Diagnostics Limited, Indian Express Building, 1 & 1/2, Dr Ambedkar Veedhi, Vasanth Nagar, Bengaluru, Karnataka 560001, India

[2] Department of Surgery, PD Hinduja Sindhi Hospital, Bengaluru, India

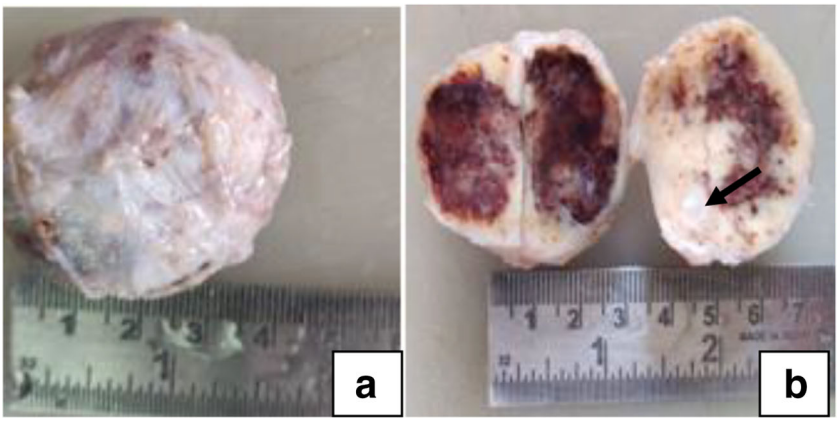

Fig. 1 **a** Gross picture showing a nodular encapsulated gray brown soft tissue. **b** Cut surface shows hemorrhage with solid tan white areas and glistening foci of the cartilage (arrow)

myoepithelioma and extraskeletal chondrosarcoma, immunohistochemistry with Pan CK and S-100 were done which showed cytoplasmic/membranous and nuclear positivity respectively in the lesional cells favoring the diagnosis of subcutaneous myoepithelioma as cytokeratins are usually negative in the latter (Fig. 3).

Discussion

Minssen first used the term mixed tumor in 1874 to describe tumors of the parotid gland with both epithelial and mesenchymal features [2]. Myoepithelial neoplasms arising in soft tissues/skin have been increasingly recognized over the last 10–15 years characterized by morphologic, immunohistochemical, and genetic means. To date, fewer than 50 cases have been reported in the world literature in various anatomic locations like the breast, larynx, soft tissues, and salivary glands [3, 4]. Cutaneous myoepitheliomas account for < 1% of all cutaneous soft tissue tumors. Only a few case reports of myoepithelioma, primarily occurring in the dermis have been described, of which < 10 cases have been reported in the foot and ankle literature. Almost 80% of cutaneous and soft tissue myoepitheliomas behave in a benign fashion with 3.4% having a risk for local

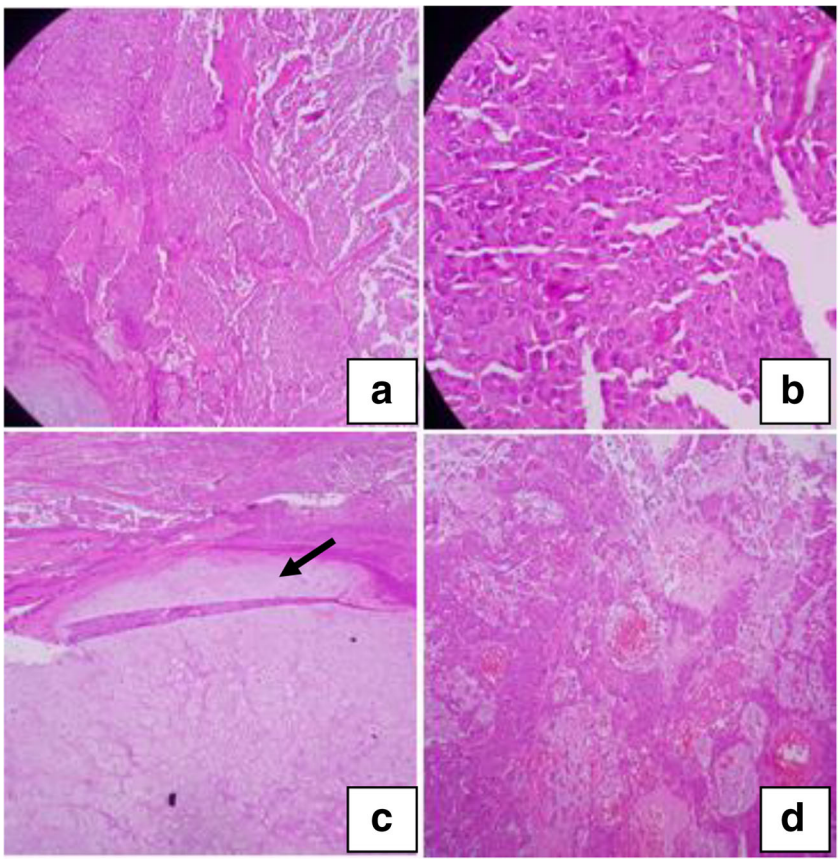

Fig. 2 **a** Tumor cells arranged in sheets, nests divided by thick fibrous septa (10 ×). **b** Plasmacytoid appearance of tumor cells with vesicular nuclei and mild to moderate nuclear pleomorphism (40 ×). **c** Foci of mature benign cartilage (arrow) (10 ×). **d** Foci of ischemic necrosis with hemorrhage (10 ×)

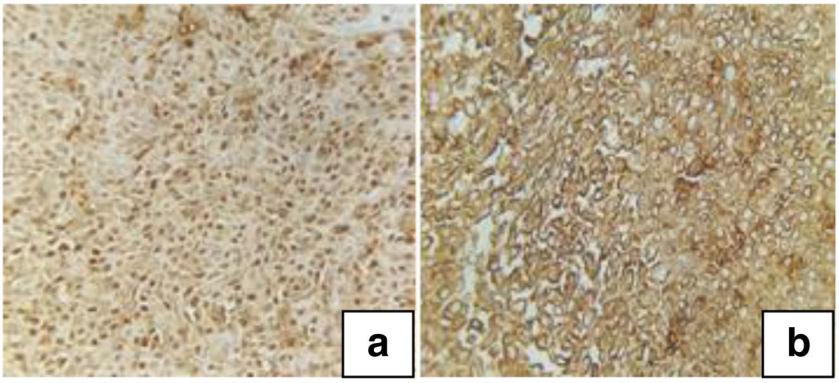

Fig. 3 **a** Diffuse nuclear positivity for S100 (10 ×). **b** Diffuse membranous and cytoplasmic positivity for PAN CK (10 ×)

recurrence and 1% having a risk of malignant transformation [5]. The incidence of these tumors ranges between 2 and 83 years of age with a mean age of presentation being 35 years with male preponderance [6]. Hornick and Fletcher published a study of 14 cutaneous myoepitheliomas of which 11 were males and 3 were females. The study indicated that cutaneous myoepitheliomas occur with peaks in childhood and in middle age, and are more commonly seen on the extremities, in contrast to the tumors of the skin, which typically occur in the head and neck region of middle-aged or elderly adults [1]. This case was unusual in its presentation in a 70-year-old male over foot region which has rarely been described in literature [4]. Most of the ankle tumors (60–70%) are benign in nature and usually include deep fibromatosis, hemangiomas, neurogenic tumors, and lipomas [7]. In the present case, initial workup with the Doppler study suggested hemangioma but a further detailed imaging study with MRI considered extraskeletal chondrosarcoma, synovial sarcoma, and nerve sheath tumors as possible differentials. Myoepitheliomas are rare and underreported tumors as they often show intratumoral heterogeneity with areas of variable cellularity, architecture, and cytologic features. These tumors are characterized by lobular growth of spindled, ovoid to epithelioid cells in reticular, trabecular, nested, and solid patterns, with a myxoid and/or hyalinized or chondroid stroma [1, 5] Other morphologic appearances include tumor cells with copious vacuolated cytoplasm (formerly so-called parachordoma), rhabdoid morphology, and plasmacytoid cells with densely eosinophilic cytoplasm as seen in the present case which further adds on the rarity of this case [8]. Heterologous differentiation can occur in up to 15% of cases (most frequently cartilaginous and/or osseous and less commonly squamous or adipocytic [2, 8]. In the present case, cartilaginous differentiation noted mislead to a potential diagnosis of extraskeletal chondrosarcoma. But the histological morphology pointed more in favor of a benign lesion as there were no mitoses, necrosis, and atypia. Considering its benign nature and close proximity to the skin with the lesion arising in the dermis, a dermal adnexal tumor, chondroid syringoma was considered as close differential along with subcutaneous myoepithelioma. Due to the wide morphologic range of myoepitheliomas, confirmatory immunohistochemistry is required to conclude at a diagnosis. As in myoepitheliomas of the salivary glands, soft tissue counterparts are positive for cytokeratins and S-100 protein [4]. A similar pattern was noted in our case where S100 showed diffuse nuclear positivity along with membranous and cytoplasmic positivity for Pan CK. The antigens most commonly expressed and therefore most sensitive for myoepitheliomas of soft tissues include a broad range of cytokeratins (expressed in nearly 100% of cases), S100 (87%), and calponin (86%) [5]. Based on morphological features coupled with the immunoprofile, the tumor was diagnosed as a subcutaneous myoepithelioma of the foot. Post-excision, the patient has been on follow-up and asymptomatic. The present case emphasizes the importance of entertaining this entity as a possible differential diagnosis in foot and ankle lesions wherein a primary diagnosis of hemangioma was considered which later was speculated to be extraskeletal chondrosarcoma on imaging as well as morphology due to the presence of heterologous elements like cartilage but finally concluded as subcutaneous myoepithelioma.

Conclusion

Subcutaneous myoepitheliomas are rare dermal tumors which should be considered as a differential diagnosis while evaluating foot and ankle lesions especially in long-standing cases where intralesional hemorrhage within the tumor mimicked hemangioma on imaging initially, surprisingly turned out to be a rare tumor of the foot. Correlation with clinical details, imaging findings, and histopathological evaluation with supporting immunohistochemistry forms a crucial part in clinching such rare diagnosis.

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