# Soft tissue mixed tumor of the hand

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## Abstract

Mixed tumors are relatively common in the skin and salivary glands, but extremely rare in soft tissues, often resulting in diagnostic problems. The occurrence of these tumors in the hand is especially limited. In this article we report the clinical, radiological, and histological features of a mixed tumor of the hypothenar region of the right hand.

## Introduction

Mixed tumors of the soft tissues were recognized as a separate entity only recently, and there is a limited number of case reports to date. Mixed tumors, along with myoepitheliomas, which constitute myoepithelial tumors, are composed of myoepithelial cells predominantly and they are relatively common in major and minor salivary glands. Because fewer than 25 soft tissue mixed tumors have been reported so far, their characterization has been scarce.[1,2] To our knowledge, there has not been a report of a mixed tumor of the soft tissue of the hand. In this report, we present a rare case of a soft tissue mixed tumor of the hand.

## Case Report

A 79-year old woman presented with a slow-growing mass of the hypothenar region of her right hand with a history of several years. There was no particular incidence of trauma or any relevant medical history. The tumor measured 4×4 cm in size and the overlying skin was smooth and nonadherent with slight redness. The patient did not complain of any pain or tenderness. Physical examination of the cervical and axillary region showed no lymphadenopathy.

On plain radiography, there was soft tissue enlargement compatible with the physical examination, but there was no calcification or bone erosion. On magnetic resonance imaging (MRI), there was a 4×4×2 cm circumscribed mass in the subcutaneous region. T1- and T2-weighted images depicted a heterogeneous lesion with intermediate intensity, slightly higher than in the muscle (Figure 1 A, B).

Incisional biopsy was performed followed by marginal resection. The tumor was well encapsulated with slight adhesion to the palmar aponeurosis (Figure 2 A, B).

On histopathology, the tumor consisted of a circumscribed lesion with a yellowish-tan appearance grossly. Hematoxylin and eosin staining demonstrated a lobulated architecture, composed of epithelioid cells and myoepithelial elements in the chondromyxoid and collagenous stroma. There was adipocytic differentiation with evidence of ductal differentiation (Figure 3A, B). Nuclear atypia was mild, and although there were sporadic mitoses, atypical mitotic figures were not identified. Immunohistochemical examination demonstrated positivity for cytokeratin, S-100, and CD10. Cytokeratin and CD 10 were positive in both spindle and epithelioid cells. S-100 protein was focally positive in the spindle cells (Figure 3C, D, E).

The postoperative period was uneventful. By her 18-month follow-up, no recurrence or metastasis had developed.

## Discussion

Mixed tumors and myoepitheliomas are well characterized in salivary glands, but were recognized to occur in soft tissue only recently.[2,3] Histological patterns are analogous to that of the salivary gland counterpart. The tumors show a spectrum of cellular and architectural morphologies. Myoepithelial cells may be spindled, plasmacytoid, ovoid, or epithelioid, and can express a wide variety of cytoplasmic filaments. The growth pattern may be solid, myxoid, or reticular, and each individual tumor includes areas with more than one growth pattern or cell subtype.[4,5]

Myoepithelial tumors of soft tissue occur equally in male and female patients. They have been reported in a wide age range with a peak in the third to fifth decades, most commonly occurring in the limbs and the limb girdles.[2,5] The vast majority of cases arise in the subcutaneous or deep subfascial soft tissue. Most patients present with painless swelling ranging in duration from a few weeks to several

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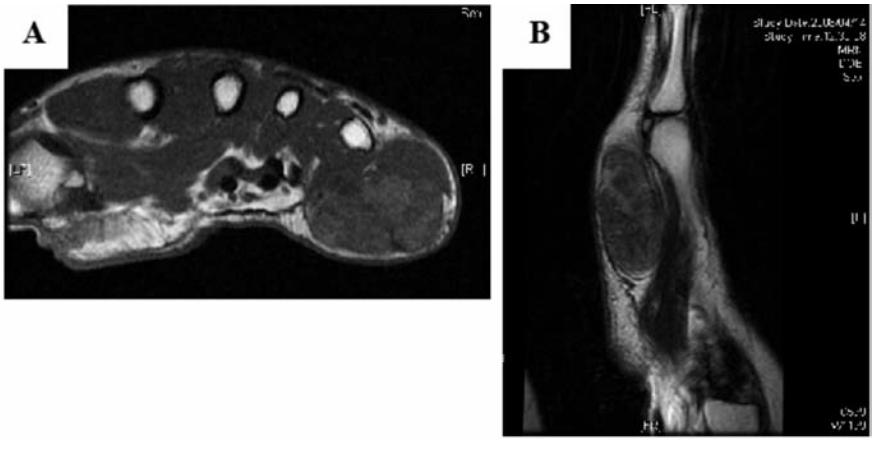


**Figure 1. (A) Axial T1-weighted magnetic resonance imaging scan revealing a 4×4×2 cm intermediate intensity mass in the subcutaneous region of the right hand. (B) Sagittal T2-weighted image showing a similar heterogeneous finding. The intensity was slightly higher than in the surrounding muscle.**

years.[5] MRI appearance of soft tissue mixed tumors varies in accordance with the heterogeneity of the tumor. Depending on the amount of hemorrhage, chondromyxoid and fibrous stroma, it is possible for the tumor to present various findings. Preoperative diagnosis based on the MRI should be performed prudently, and incisional biopsy should be performed at all times.

There is a debate still as to whether mixed tumors and myoepithelioma should be distinguished as separate entities or considered as the same spectrum of tumors with overlapping histological appearances and similar clinical behavior. Those tumors either lacking or with very limited ductal differentiation generally are classified as myoepitheliomas. Current classification simply separates those tumors with ductal differentiation into the mixed tumor categories.[6,7] Whereas some investigators allow up to five percent or ten percent ductal differentiation in myoepitheliomas,[4,8-10] others classify tumors with any ducts as mixed tumors. In the present case, the tumor showed distinct duct formations; therefore we diagnosed it in accordance with the strict criteria.

The absence of clear-cut histopathological clues for the diagnosis of myoepithelial tumors is hampered further by the wide variability in their immunohistochemical characteristics. Immunoreactivity for the S-100 protein and muscle actins seems to be the most constant immunophenotype, whereas immunoexpression for epithelial markers such as cytokeratins and EMA, or neural markers such as GFAP, is somewhat erratic and variable from case to case. The same immunophenotype has been described in salivary gland myoepithelial tumors, which usually coexpresses immunoreactivity for S-100 protein, muscle actins, and GFAP, with variable immunoexpression for EMA and cytokeratins.[8,11-16]

Although the majority of morphologically benign-looking mixed tumors of soft tissue behave in a benign fashion, there is approximately a twenty percent risk for local recurrence. The malignant potency of myoepithelial tumors varies, and it is difficult to differentiate myoepithelial tumors into benign and malignant categories on histological grounds only.[17,18] For the diagnosis of malignant myoepithelioma in salivary glands, an invasive growth pattern has been considered as the most important feature, because the immunohistochemical features are similar for the benign and malignant forms.[18] In addition, cytological atypia and mitotic rate have been reported to be useful.[19,20] However, in the case of soft tissue myoepithelial tumors, there has been no association between the degree of nuclear pleomorphism or mitotic activity and clinical behavior.[21] Recurrent chromosome rearrangements, particularly reciprocal translocations, with breakpoints on 8q12, 3p21, and 12q14-15 have been described in myoepithelial tumors of the salivary gland.[22] It is interesting to note that the malignant myoepithelial tumor has been reported to show different chromosomal abnormalities such as gains of 1p31~p34, 1q21~q23, and 16q22, and loss of 15q.[23] Nevertheless, it is difficult to establish prognostic indicators for soft tissue myoepithelial tumors until further reports are available.

Because there is considerable morphologic heterogeneity of soft tissue myoepithelial tumors, the differential diagnosis should be based on the dominant histological pattern. If the tumor displays a reticular architecture with chondromyxoid or hyalinized stroma, extraskeletal myxoid chondrosarcoma and ossifying fibromyxoid tumor should be considered as differential diagnoses. Solid spindle cell myoepithelial tumors can resemble leiomyomas and schwannomas. Furthermore, if the tumor shows significant cytological atypia, metastatic carcinomas, metastatic melanomas, and epithelioid sarcomas should be considered as well.

The mixed tumor should be considered as one of the differential diagnoses of soft tissue tumors of the hand. The rarity of this tumor has not enabled prediction of the possible out-

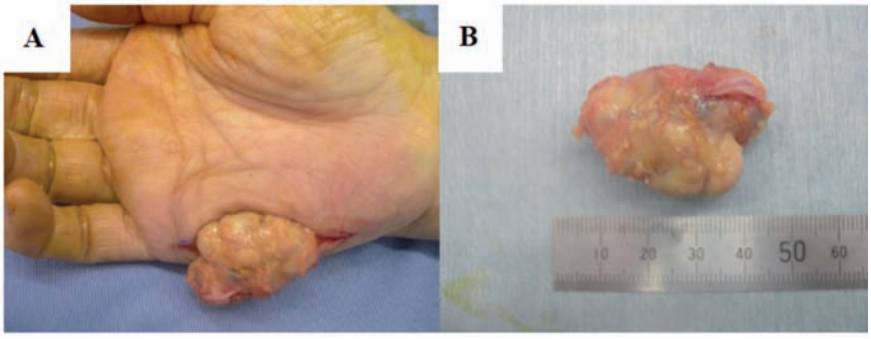


**Figure 2. (A, B) A 4×4×2 cm soft tissue mixed tumor of the hypothenar region of the hand.**

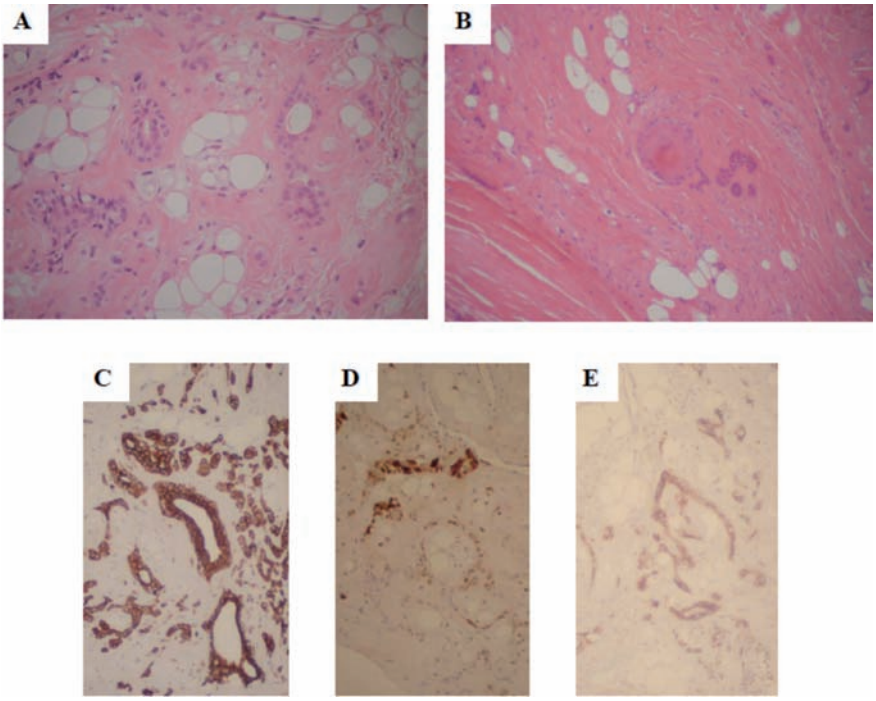


**Figure 3. (A, B) Photomicrograph of the soft tissue mixed tumor, showing a lobulated architecture with epithelioid cells and myoepithelial elements in the chondromyxoid and collagenous stroma, and evidence of ductal differentiation (H&E stain). Immunohistochemical examination demonstrated positivity for cytokeratin (C) and CD10 (E) in both spindle and epithelioid cells, and S-100 protein in the spindle cells (D).**

come after resection. Because there are reports of local recurrence and malignant transformation, complete resection with appropriate follow-up of the patients should be warranted.

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