

# Myoepithelioma of the Hand and Carpal Tunnel: An Unusual Cause of Median Nerve Compression

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**ABSTRACT:** Myoepitheliomas are rare tumors increasingly recognized to occur in the soft tissues. Although the hand and carpal tunnel are exceptional locations, the presence of these lesions in such sites constitutes a potential cause of debilitating symptoms. We report the case of a patient with severe pain secondary to median nerve compression and displacement of flexor tendons caused by a rapidly growing myoepithelioma. This is the first sonographic description of this tumor producing carpal tunnel syndrome and disabling pain. High-resolution sonography allowed evaluation of gross tumor morphology and real-time assessment of its interactions with surrounding structures. © 2010 Wiley Periodicals, Inc. *J Clin Ultrasound* 39:44–47, 2011; Published online in Wiley Online Library (wileyonlinelibrary.com). DOI: 10.1002/jcu.20740

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## INTRODUCTION

Myoepitheliomas had been mostly described in salivary glands until their more recent recognition in soft tissues.<sup>1</sup> These tumors display a wide range of pathologic and morphologic characteristics,<sup>2</sup> which, in addition to a lack of well-

defined diagnostic criteria, has complicated their identification and classification.

The hand and, in particular, the carpal tunnel, are exceptional locations for myoepitheliomas. So far, we have only found two reports of such tumors causing unilateral carpal tunnel syndrome.<sup>3,4</sup> Herein, we report the case of a patient with median nerve compression caused by a rapidly growing myoepithelioma. To the best of our knowledge, this is the first sonographic documentation of this tumor producing median nerve compression and carpal tunnel syndrome with debilitating pain.

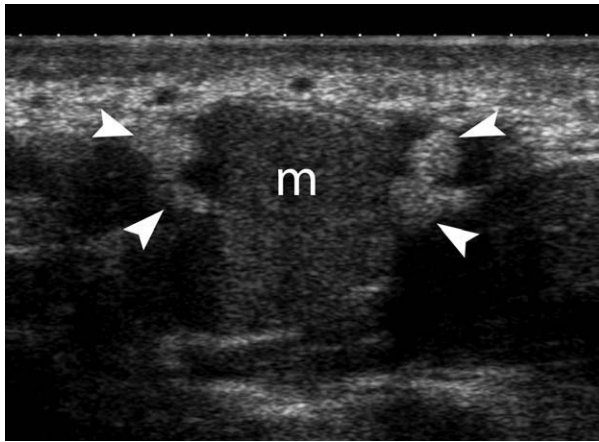
## CASE REPORT

A 35-year-old female patient with no significant medical history presented with a mass in the anterior aspect of her left hand and wrist. She initially complained of minimal discomfort, which progressed to severe pain as the lesion grew over a period of 3 months. She also noted slight paresthesias along the distribution of the median nerve with weakness and some limitation of palmar flexion. On clinical examination, the mass was soft, relatively mobile, and mildly tender.

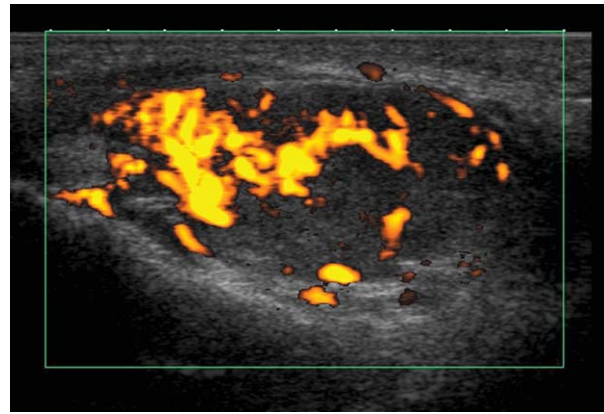
Sonographic examination with a Aplio SSA-770A scanner equipped with linear multifrequency transducers (9–12 and 9–14 MHz) (Toshiba Medical Systems, Tokyo, Japan) revealed a 5 × 2

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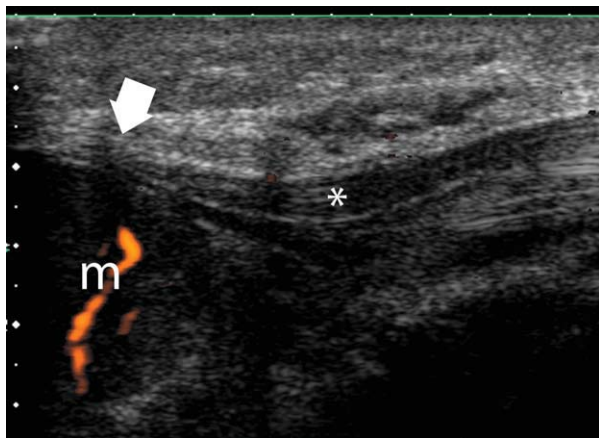
## MYOEPITHELIOMA OF THE HAND AND CARPAL TUNNEL



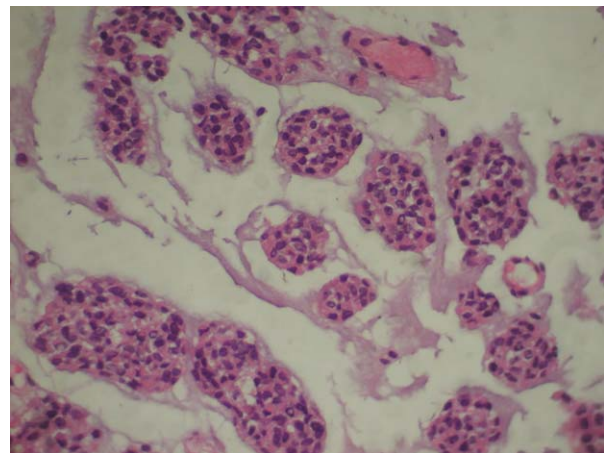
**FIGURE 1.** Short-axis sonogram immediately below the carpal tunnel outlet shows a solid, well-defined, homogeneous, lobulated, and hypoechoic mass (m) between the flexor digitorum tendons of the index and middle fingers (arrowheads).



**FIGURE 3.** Power Doppler sonogram shows the hypervascular mass.



**FIGURE 2.** Longitudinal sonogram of the median nerve at the level of the distal carpal tunnel demonstrates a hypochoic palmar mass (m) that causes anterior displacement and compression (arrow) of the median nerve (\*).



**FIGURE 4.** Photomicrograph shows a myxoid-type stroma surrounding clumps of cells arranged in an organoid and trabecular pattern. There is no evidence of mitotic activity, hemorrhage, or necrosis. No glandular structures are seen (H&E stain;  $\times 40$ ).

$\times 1.6$  cm, well-defined, homogeneous, lobulated hypochoic mass overlying the proximal second and third metacarpal bones and extending cephalad into the distal carpal tunnel, producing splaying of the flexor tendons (Figure 1). The superior pole caused anterior displacement and compression of the median nerve (Figure 2), which appeared enlarged within the carpal tunnel (cross-sectional area,  $0.13 \text{ cm}^2$  at inlet and  $0.14 \text{ cm}^2$  at outlet). The lesion showed increased blood flow on power Doppler imaging (Figure 3). Once the tumor morphology and its relations with surrounding structures were clearly described by sonography, and especially given the patient's intense pain and rapid tumor growth, it was decided to proceed with surgery without further imaging. The patient underwent surgical excision of a soft, lobulated,

and spongy mass. Attachment to the median nerve accounted for a lengthy and complicated dissection. Pathologic analysis showed a myxoid-type stroma surrounding trabecular sheets of cells with an organoid pattern. Nuclei were round to oval, some featuring clumped chromatin and irregular borders. There was no evidence of mitotic activity, hemorrhage, or necrosis (Figure 4). Immunohistochemical studies showed some immunoreactivity with antibodies to epithelial membrane antigen and focal immunoreactivity with S-100 protein. Glial acidic fibrillary protein immunostain was negative and keratin (AE1/AE3) stain was for the most part negative with only questionable foci of immunoreactivity. Although an extraskeletal myxoid chondrosarcoma was in the differential diagnosis, the final histologic and immunohistochemical findings were those of a soft tissue myoepithelioma.

There was no clinical or sonographic evidence of recurrence at 6 months and 2 years after surgi-

cal excision. The patient still has some residual weakness, mainly on the index finger, but has progressively regained most function on her left hand and denies pain.

## DISCUSSION

Myoepithelial cell tumors show similar gender distribution and a peak incidence between the third and fifth decades.<sup>2</sup> They can present in pure form (myoepitheliomas or myoepithelial carcinomas) or be associated with glandular structures (mixed tumors). Most myoepitheliomas have been reported in the salivary glands,<sup>5</sup> with fewer cases in different parts of the body. Their recognition in soft tissues is relatively recent,<sup>1</sup> perhaps in part due to their low frequency as well as a wide range of morphologic and histologic features.<sup>2,5,6</sup> This has resulted in unclear and sometimes conflicting diagnostic criteria.

The majority of soft tissue myoepitheliomas are found in the extremities and limb girdles, followed by head and neck, and trunk.<sup>2</sup> Their localization in the hand or wrist is unusual and these tumors generally do not cause a mass effect or compressive symptoms. After review of the medical literature through PubMed (separate queries using the keyword "myoepithelioma" paired with "hand," "soft tissues," and "carpal tunnel"), we found only two cases of myoepithelioma within the carpal tunnel producing compression of the median nerve.<sup>3,4</sup> In the largest series on soft tissue myoepitheliomas, only one case showed non-specified paresthesias while more than half of the patients had a painful mass.<sup>2</sup> Our patient's symptoms were so severe that she had trouble with grasp function and was continuously dropping objects to the floor. Such an aggressive clinical presentation might be explained by rapid growth of the tumor expanding into a small compartment such as the carpal tunnel.

Criteria for malignancy have not been well established, particularly in soft tissues. Capsule invasion might be helpful when these tumors are located in the salivary glands, but this obviously cannot be directly applied to a soft tissue context. We also speculate that certain features such as pattern of infiltration may differ depending on the location of the lesion (superficial versus deep tissues, restraining compartments versus relatively ample spaces, etc). In the series by Hornick and Fletcher, 18% of tumors classified as having mild or no atypia showed recurrence, while tumors with clearly malignant cytology showed rates of local recurrence and metastasis of 42% and 32%, respectively.<sup>2</sup> The two myoepithelial

tumors previously reported in the carpal tunnel showed mild atypia and no signs of recurrence; follow-up, however, was short (5 months in both reports). Despite the fact that we found no atypical or otherwise malignant microscopic features in our case, we were certainly concerned about recurrence, given the initial rapid growth of the lesion. Fortunately, follow-up at 2 years showed no signs of recurrence.

Although imaging findings in reported cases are nonspecific, features such as lobulation, well-defined margins, and avid uptake of contrast material appear to be common. MR shows lesions that are hypo-, iso-, or hyperintense to muscle in T1-weighted images,<sup>7-9</sup> sometimes with areas suggestive of hemorrhage.<sup>8</sup> The tumors appear hyperintense and are often heterogeneous on T2-weighted imaging and may also have septa.<sup>4,7-9</sup> MR with gadolinium shows diffuse uptake that is mostly inhomogeneous,<sup>9,10</sup> which is similar to data from contrast-enhanced CT studies.<sup>7,11</sup> This is consistent with the tumor in our report, which was hypervascular by Doppler assessment. We also found one instance of thallium-201 uptake with complete subsequent washout.<sup>4</sup> Interestingly, another case showed positive fluorine-18-fluorodeoxyglucose uptake on positron emission tomography imaging in a lesion that proved to be benign by histologic analysis.<sup>9</sup> The limited sonographic descriptions of myoepithelial tumors in other body parts report various types of echogenicity. One salivary myoepithelioma arising in a masseter muscle was described as a solid and hypoechoic mass with irregular margins, internal inhomogeneities, and a hilar vascular pattern.<sup>7</sup> A separate report of a myoepithelioma of the breast described the mass as intermediately echogenic, solid, and slightly lobulated, with ill-defined borders and some posterior shadowing. Doppler vascularity was not documented.<sup>10</sup> The mass in our case was lobulated, well-defined, homogeneously hypoechoic, and hypervascular.

The sonographic anatomy of the wrist, including the relationships of the median nerve to surrounding bony structures and flexor tendons, has been previously documented.<sup>12</sup> Quantitative evaluation of median nerve cross-sectional areas has proven to be accurate and highly predictive of carpal tunnel syndrome.<sup>13,14</sup> Interestingly, the median nerve in our patient showed notable enlargement along its course throughout the tunnel, an effect that was produced by a mass largely located in the hand and involving a relatively small section of the distal carpal tunnel.

In summary, the paucity of the cases precludes an analysis of typical imaging features. However,

it is reasonable to expect that a wide histomorphologic spectrum would translate into non-specific imaging characteristics. In our case of a particularly symptomatic and functionally impaired patient, high-resolution sonography was effective in evaluating the gross tumor morphology and, most importantly, in determining in real-time its relationship with adjacent nerves and tendons.

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