Myopericytoma of the hypothenar eminence: case report

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Abstract Myopericytomas belong to a benign group of tumors consisting of perivascular myopericytes or pericytes with a characteristic myoid differentiation pattern. The tumor is histologically and radiographically heterogenous, similar to superficial myofibromas. We report on a case of a myopericytoma arising in the hypothenar eminence of a 58-year-old man with a history of over 15 years of ulnar-sided and midcarpal wrist pain. Decision was made to excise the mass after it had increased in size by over 50 % over a 10-year period. Excision of the mass led to remarkable relief of his symptoms in the hypothenar as well as midcarpal regions. Consideration of this rare perivascular tumor should be included in patients with slow-growing benign appearing masses within the subcutaneous tissues of the hand.

Introduction

Myopericytomas are a group of perivascular tumors consisting of myoid cells [3]. Usually located in the subcutaneous or dermal tissues, it displays a range of histologic growth patterns [2]. Initially described in 1996 as a subset of cutaneous myofibromas consisting of proliferating myocytes, this term has been expanded to include tumor consisting of perivascular proliferation of spindle cells with a myoid differentiation [6, 8]. In 2002, the World Health Organization

(WHO) recognized the term myopericytoma to describe these perivascular tumors with myoid differentiation. These tumors usually not only involve the skin, vessels, or soft tissues in the lower extremities, but can also involve the head, neck, axial skeleton, or upper extremity [2, 7]. Although usually benign, there have been cases of malignant myopericytomas [6]. Although it has been described in the arm, the hand is a relatively rare location for myopericytomas to occur. One case involved an intravascular tumor involving the superficial palmar arch [10]. Another occurrence of myopericytoma in the hand was described in a woman with multiple cutaneous nodules throughout the body including the hand [7]. In both cases, the tumor presented as a slow-growing perivascular mass that lead to persistent discomfort without any preceding trauma.

The purpose of this article is to report an unusual presentation of this rare tumor and provide a review of the available literature.

Case Description

A 58-year-old right-hand dominant man with a chronic history of ulnar-sided right wrist pain was evaluated for a hypothenar mass. The pain had been present for over 15 years, localized to the ulnar wrist, as well as the hypothenar eminence near the pisiform and hook of the hamate, with occasional median and ulnar nerve numbness and paresthesias. Although he had a history of several wrist sprains from hyperextension injuries, he did not have any identifiable trauma preceding the onset of the pain. Physical examination revealed normal grip, appositional and oppositional pinch strength, but slightly increased 2-point discrimination in the ulnar nerve distribution. Deep palpation in the area of the hook of the hamate reproduced the pain and paresthesias.

Investigation performed at the Mayo Clinic, Rochester, MN

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Methods

CT scan obtained 10 years prior to his most recent evaluation was negative for a hook of the hamate fracture. EMG was demonstrated bilateral carpal tunnel syndrome. Remarkably, MRI revealed a small $5 \times 5 \times 5$ -mm heterogenous mass with subtle enhancement on T2 imaging in the area of the hypothenar eminence, along with midcarpal degenerative and first carpometacarpal joint changes (Fig. 1). Given his midcarpal pathology and carpal tunnel syndrome, the decision was made to undergo surgical management. The patient subsequently underwent a carpal tunnel release, anterior and posterior interosseous neurectomies, and arthroscopy of the radiocarpal and midcarpal joints. Arthroscopy revealed degenerative changes of his TFCC, with arthritic changes of the proximal pole of the hamate.

The midcarpal pain gradually returned after the procedure localized to the capitolunate articulation dorsally. The patient received numerous midcarpal corticosteroid injections that were effective for pain relieve of his midcarpal arthritis. Over the course of 10 years, the patient's pain localized to the region of the hook of the hamate, with the sensation of a soft tissue mass rubbing against it. The pain and paresthesias were only able to be reproduced with deep palpation of hypothenar eminence. Ultrasound demonstrated a $8 \times 10 \times 6$ -mm hypoechoic nodule in the hypothenar eminence, increasing in size from the MRI performed 10 years prior (Fig. 1). The mass had no relation to the ulnar artery or nerve. The ultrasound and prior MRI were consistent with a benign tumor, with the differential including peripheral nerve sheath tumor, glomus tumor, or angioleiomyoma.

The patient underwent open excision of the hypothenar mass using a Bruner-type incision directly over the mass. The mass was smooth, spherical, and well-circumscribed, located superficial to the ulnar nerve and artery (not in continuity with) in the hypothenar adipose tissue (Fig. 2). Pathology revealed a benign myopericytoma (Fig. 3). The patient reported that he experienced immediate pain relief in both the hypothenar and midcarpal regions, despite the presence of his midcarpal degenerative changes.

All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008. Informed consent was obtained from all patients for being included in this study.

Discussion

The term myopericytoma was first described when Dictor et al. described a myofibromatosis-like hemangiopericytoma in a young boy [1]. Pericytes are thought to be pluripotent cells, capable of differentiating into multiple lineages, such as smooth muscle vascular cells. The myopericyte is a precursor cell to these differentiated smooth muscle cells [7]. Thus, when these cells fail to terminally differentiate and instead increase their rate of proliferation, myopericytomas begin to form. Their histologic and gross appearance is somewhat heterogeneous, often resembling multiple cutaneous and subcutaneous entities, such as hemangiopericytoma, angioleiomyoma, or more commonly myofibromas, and thus were originally thought to be a subset of these cutaneous

Fig. 1 Tumor imaging. Ultrasound (*top left*) revealed a $8 \times 10 \times 6$ -mm hypoechoic nodule in the hypothenar eminence, increasing in size from the MRI performed 10 years prior, which demonstrated a small $5 \times 5 \times 5$ -mm heterogenous mass with subtle enhancement on T2 imaging in the area of the hypothenar eminence

Fig. 2 Gross tumor specimen. The mass was smooth, spherical, and well-circumscribed, located superficial to the ulnar nerve and artery (not in continuity with) in the hypothenar adipose tissue

tumors [6, 8]. However, the unifying characteristic with all myopericytomas is the perivascular myoid differentiation in each tumor.

These tumors are considered slow-growing tumors with diameters generally less than 2 cm. These have been reported

anywhere in the body, but seem to have a predilection for the lower extremity [2, 3, 7]. In rare cases, these tumors can be multifocal with locations throughout the body's cutaneous or subcutaneous tissues [2–4, 7]. As was demonstrated without patient, the myopericytomas are characterized as being well-

Fig. 3 Tumor pathology. Pathology revealed a benign myopericytoma with positive staining for desmin and smooth muscle actin (SMA). As seen, the unifying characteristic with all myopericytomas is the perivascular myoid differentiation in each tumor

demarcated, heterogenous enhancing, hypoechoic masses on imaging modalities, such as MRI, ultrasound, or CT scan [4].

Although myopericytomas are common in the lower extremities and have been described in the proximal upper extremity, the hand is an extremely rare location for it to arise. Woollard et al. described a case where a benign myopericytoma arose intravascularly in the superficial arch of a 63-year-old man [10]. The gross appearance demonstrated a spherical swelling of the vessel wall, while histology revealed endothelial cells surrounded by undifferentiated smooth muscle cells. One of the cases described by Mentzel involved a 20-year-old woman with painful cutaneous nodules of the hand, forearm, thigh, calf, foot, and vulva [7]. Most of the lesions were confined to the dermis and associated with subcutaneous vessels. All lesions were excised except the lesion on the vulva, without any local recurrence or metastasis. Unlike the location in our patient, the myopericytomas in this and other series appear to be localized to the skin and subcutaneous vasculature of the hand [2, 7]. However, similar to our patient, these tumors appeared as benign, slow-growing masses, leading to persistent discomfort without any preceding trauma.

Often, myopericytomas are associated with blood vessels. Some reports suggest that they are similar in gross cytologic appearance to glomus tumors [2, 7]. However, they are differentiated from them by the elongated spindle cell pattern and presence of undifferentiated myopericytes. Although usually perivascular in location, there have been numerous reports of intravascular locations [2, 5, 7, 9, 10]. McMenamin et al. described a case of an intravascular tumor within a vein in the thigh [5]. Although usually these tumors form spontaneously without a history of trauma, Scott et al. described a myopericytoma arising intravascularly in leg vasculature after a history of repeated trauma [9]. Interestingly, the myopericytoma in our patient did not seem to be localized to or arise from any blood vessel. Instead, it seemed to arise from the adipose tissue within the hypothenar eminence.

These tumors are usually benign in nature. They are usually treated successfully with complete surgical excision. However, they have been known to recur after local excision. Furthermore, there have been reports of malignant pericytomas leading to distant metastases. McMenamin et al. described five malignant pericytomas involving the upper arm, neck, and lower extremity [5]. All four patients with deep myopericytomas developed metastases and three died within 1 year of presentation, while the patient with a superficial location remained metastasis free at 18 months of follow-up. One of the tumors in Mentzel's series of myopericytomas fit the histologic and morphologic criteria for malignancy [7]. This superficial tumor recurred locally after excision, but did not metastasize to any distant locations.

This case presents a rare myopericytoma arising in the hypothenar eminence without any obvious association with a blood vessel. The tumor was slow growing, increasing in size by just over 50 % over a 10-year period. The primary discomfort from local compression on the hook of the hamate eventually led to excision of the mass and diagnosis of myopericytoma. Although the patient did demonstrate midcarpal arthritis, excision of this mass led to significant relief of not only his hypothenar but also midcarpal pain.

Disclosures Each author certifies that his or her institution approved the human protocol for this investigation and that all investigations were conducted in conformity with ethical principles of research.

Conflict of Interest Eric R. Wagner declares that he has no conflict of interest.

Alexander Y. Shin declares that he has no conflict of interest.

Statement of Human and Animal Rights IRB approval was obtained.

Statement of Informed Consent Informed consent is not applicable since this is a retrospective review.

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