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Painful Palmar Mass in a 36-year-old Woman

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History and Physical Examination

A 36-year-old right-hand-dominant woman presented with a 2-month history of intermittent sharp stabbing pain in the left palm. She denied any history of distinct trauma or infection in the area. She was active as a college Frisbee coach and played golf recreationally. Severe pain occurred only when playing Frisbee or golf or during handshaking. The patient denied pain at night. On inspection of the hand, there was no visible mass or deformity. Physical examination of the hand revealed firmness in the palm at the base of the third metacarpal. This area was locally tender to palpation, and pressure applied to this area reproduced the patient's painful symptoms. There were no skin changes overlying the affected area. Motor and sensory function were preserved in median, radial, and ulnar nerves. Two-point discrimination was preserved distally at the fingertips. The patient had full range of motion at the wrist and in the

metacarpophalangeal joints and interphalangeal joints of all digits. Tinel's sign was negative at the wrist, and Allen's test revealed patency of the radial and ulnar arteries.

We initially obtained plain radiographs of the affected hand. These films were interpreted as normal. Subsequently, MRI was performed to better characterize the location and nature of the mass (Figs. 1, 2).

Based on the history, physical examination, and imaging studies, what is the differential diagnosis?

Imaging Interpretation

T1-weighted MR images showed a well-defined 5-mm rounded mass lying just deep to the palmar fascia and superficial to the flexor tendons at middiaphyseal level between the index and long metacarpals. The lesion abutted but did not envelope or substantially displace the adjacent flexor tendons. The lesion was isointense to muscle (Fig. 1). Further sequencing revealed the mass to be homogeneous and hyperintense on T2-weighted imaging, suggesting the possibility of a fluid component to the mass. No surrounding edema was noted on the T2-weighted images (Fig. 2). No contrast was administered.

Differential Diagnosis

Dupuytren's nodule
Ganglion cyst
Fibroma of tendon sheath
Perivascular tumor (hemangiopericytoma, angioleiomyoma, glomus tumor, myopericytoma)
Giant cell tumor of tendon sheath
Neurilemmoma (Schwannoma)

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Each author certifies that his or her institution has approved the reporting of this case report and that all investigations were conducted in conformity with ethical principles of research.

This work was performed at Wake Forest University Baptist Medical Center.

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Fig. 1 A T1-weighted MR image reveals a 5-mm mass (arrow) isointense to muscle. The mass is located in the middiaphyseal region between index and long metacarpals, deep to the palmar fascia and superficial to the flexor tendons.

Fig. 2 The mass (arrow) is hyperintense on T2-weighted MRI and again is deep to the palmar fascia but superficial to the flexor tendons in the palm.

Owing to the small size of the lesion, an excisional biopsy was performed. Intraoperatively, a 5-mm-diameter smooth-surfaced nodule was found arising from a small branch of the superficial palmar arch. The nodule was lying between the superficial arch and common digital nerves along the third metacarpal (Fig. 3). The mass was dissected off the surrounding soft tissue and vessels (Fig. 4). Intraoperative frozen section was not obtained. The specimen was sent to pathology for histologic examination.

Based on the history, physical examination, imaging studies, and histologic studies, what is the diagnosis and how should this lesion be treated?

Histology Interpretation

Low-power histologic examination revealed an encapsulated tumor composed of bland spindle cells with multiple small, scattered vascular spaces (Fig. 5). High-power histologic review revealed concentric spindle cell ringing the

Fig. 3 After dissection through the palmar fascia, the mass was seen at the level of the superficial palmar arch.

Fig. 4 The mass was dissected from the surrounding soft tissue; it arose from a small arterial branch off the superficial palmar arch.

Fig. 5 A low-power microscopic view of the tumor shows encapsulation and multiple scattered vascular spaces in a background of bland spindle cells (Stain, hematoxylin and eosin; original magnification, $\times 100$).

Fig. 6 A high-power microscopic view reveals concentric spindle cell ringing of multiple small vascular spaces (Stain, hematoxylin and eosin; original magnification, $\times 200$).

vascular spaces (Fig. 6). The spindle cells were positive for α -smooth muscle actin on immunohistochemical staining.

Diagnosis

Myopericytoma

Discussion and Treatment

A complaint of a painful nodule in the palm may produce a broad differential diagnosis. However, certain findings help to exclude some diagnoses while suggesting others. Pertinent findings in this patient included lack of overlying skin changes, normal range of motion and neurovascular examination, and reproduction of the patient's painful symptoms with deep palpation. The radiographic findings were nonspecific; revealing a well-circumscribed lesion isointense to muscle on T1-weighted imaging. The histologic features of the lesion consistent with myopericytoma included the presence of scattered vascular spaces with concentric spindle cell ringing and a positive α -smooth muscle actin immunohistochemical stain.

The nodule in this patient was not believed consistent with Dupuytren's disease. There were no other palpable nodules, cords, or skin changes, and range of motion of the digits was normal. The location and appearance of the lesion on physical examination and MRI were not considered typical of a ganglion cyst. At the time of excisional biopsy, the differential diagnosis remained broad. Only after the lesion was observed arising from the superficial palmar arch could fibroma of tendon sheath, giant cell tumor of tendon sheath, and neurilemmoma be excluded. The exclusion of other perivascular tumors, such as

hemangiopericytoma and angioleiomyoma, was enabled only by histologic evaluation.

Myopericytoma falls into the broader category of perivascular soft tissue neoplasms. It is described only in case reports in the orthopaedic literature [1, 12] and was delineated as a distinct entity by pathologists in the 1990s [4]. Most perivascular tumors had been divided into either glomus tumors or hemangiopericytomas previously [9]. However, the term hemangiopericytoma encompasses a heterogeneous group of tumors, allowing for overlap of tumors with myoid differentiation and even some glomus tumors, as hemangiopericytoma [2]. A hemangiopericytoma-like vascular growth pattern can be found in neoplasms of differing cell lineages, ranging from fibrous tissue to chondroid tissue [6]. Consequently, the existence of the hemangiopericytoma as a distinct clinical entity was challenged [3]. The term myopericytoma, initially used to describe a subset of cutaneous myofibromas originating from the myopericyte [10], has gained support as the designation for a subset of neoplasms that would have been included in the broad hemangiopericytoma category. It was entered as a subgroup of perivascular tumors in the World Health Organization Classification of Soft Tissue Tumors [7].

Myopericytoma occurs most frequently in the distal extremities [2]. In the largest published case review, 42 of 54 cases occurred in the extremities [9]. Additionally, myopericytoma tends to arise in either the dermis or superficial subcutaneous tissues [2]. In a review of 54 cases, only four were found arising from deep soft tissues [9]. If the lesion involves the dermis, it may appear as a white nodule but also may have a purplish hue, mimicking a hemangioma [2]. Periungual involvement also has been described [11]. The lesion is typically painless unless pressure is applied [2].

The neoplasm often arises in the midadult years [2, 4, 10]. However, it may occur in young adults or children [10]. When the lesion occurs in younger patients, there is an increased likelihood of multiple lesions, often occurring in the same anatomic region [7]. Previous reports include a 13-year-old boy who had two lesions on his left leg during a 16-month period [2], and a 19-year-old man who had two lesions simultaneously on his right lower leg [9].

Radiographic characterization of the lesion is limited to one case report [5]. In this case, the mass was seen on plain radiographs as a soft tissue swelling but was unusually large at 4 cm in length. On MRI, the mass was heterogeneous but predominantly isointense to muscle on T1-weighted images and hyperintense on short-tau inversion recovery images. On postgadolinium images, the mass showed intense signal enhancement [5].

There appears to be a small potential for malignancy. In the largest series available, one neoplasm in 54 met the criteria for a malignant designation [9]. The lesion recurred

after initial marginal excision but after subsequent wide excision, no additional local recurrence or systemic metastases were present at the 3-year followup. A separate series reviewed five cases of malignant myopericytoma [8]. One was at the site of previous multiple benign myopericytomas. Four of the five tumors metastasized. The one neoplasm that did not metastasize was superficial in location. In contrast, the metastasizing tumors were all deep-seated lesions. This suggests a correlation between the depth of the lesion and the clinical course.

Although the potential for malignant transformation exists, the incidence remains very low. Consequently, myopericytoma classically is still considered a benign, slow-growing tumor with a low rate of local recurrence [2, 9]. Dray et al. [2] reported all seven cases in their series were slow-growing with a low mitotic rate, with one tumor having been present for 12 years at the time of diagnosis. There were no instances of local recurrence [2]. Mentzel et al. [9] reported only two cases of recurrence in 46 tumors at a mean followup of 48 months, despite marginal or incomplete excision in 23 of 46 cases.

Surgery for these lesions must be patient-specific based on history, location, and histologic findings. We recommend excisional biopsy of small lesions. Marginal resection is an acceptable technique, given the documented low rate of recurrence, even with incomplete excision. However, certain situations mandate a more aggressive wide resection be considered. These include a myopericytoma at the site of multiple prior benign myopericytomas, deep-seated lesions, and those with histology indicative of malignant transformation. We believe needle biopsy should be considered before excision with larger lesions and those that arise at the site of multiple prior benign lesions. We recommend yearly clinical examination as followup for patients with lesions that are superficial or palpable. Followup for patients with deeper lesions should be at the discretion of the treating surgeon. There is insufficient evidence to recommend for or against routine surveillance MRI. Malignant lesions should be evaluated for any metastatic disease [8].

Our patient was managed with marginal excision at the time of surgery. Final histologic examination confirmed a

benign process. At the 2-year followup, there was no evidence of recurrence. Our case is the second reported case of myopericytoma involving the superficial palmar arch, which could be an unrecognized favored location.

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