Digital Myopericytoma: A Case Report and Systematic Literature Review

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J Hand Microsurg 2017;9:32–36.

Abstract

A myopericytoma (MP) is an exceedingly rare perivascular tumor of unknown etiology. Given their potential for mimicry and malignancy, MP tumors pose a unique challenge for surgeons and may be overlooked on differential diagnosis. We present a case report of an otherwise healthy 33-year-old right-hand dominant male who presented to our outpatient clinic with a 2-month history of painless swelling and erythema of the pulp of his left index finger. Subsequent plain film X-ray showed near-complete bony destruction of his distal phalanx. Pathological evaluation of an incisional biopsy showed a benign variant of MP. The lesion was treated by excision with tumor shelling, and there was no evidence of recurrence 81 days postoperatively. A systematic literature review of the management and outcome of all known cases of hand and wrist MP is presented.

Keywords

- myopericytoma
- soft-tissue tumor
- case report

Introduction

A myopericytoma (MP) is a rare perivascular growth of unknown etiology. It was first classified in 1998 by Granter et al,¹ and has since been described by the World Health Organization as a soft-tissue tumor.² It falls on a morphological continuum with other lesions such as myofibroma and angioleiomyoma. Although by definition myopericytotic lesions are benign, recent reports of malignancy have been documented.³ Given their potential for mimicry, MP tumors pose a unique challenge for surgeons and may be overlooked on differential diagnosis. This case report and comprehensive literature review will provide direction for the identification and management of MP lesions presenting in the hand and wrist.

In the largest case series of MP tumors presenting in all anatomical locations, lesions were most frequently encountered in cutaneous tissues, showing a characteristic slow but steady enlargement over time.⁴ Furthermore, despite half of these lesions undergoing incomplete excision, there were only two local recurrences.⁴ The lower extremities are the most commonly affected anatomical region, followed by the upper extremities and head and neck. Although most lesions

present in isolation, there are a few documented instances of multiple primary tumors.⁵

Most cases of MP appear to be idiopathic, though there are isolated cases associated with trauma. In one small case series, lesions developed following facial abrasions at the alveolar ridge and forehead.⁶ Acquired immunodeficiency syndrome has also been linked to the development of MP.⁷ Unfortunately, both of these case series are limited by sample size.

Histopathologically, MP tumors exhibit a distinguishing cellular pattern made evident by oval-to-spindle appearance and concentric, ring-like perivascular invasion. On immunologic staining, lesions exhibit positive smooth muscle actin staining and, in certain cases, CD34 positivity.² Several different histological subtypes have also been described, and include the following: solid MP, hemangiopericytoma-like MP, angioleiomyoma-like MP, myofibroma-like MP, hypocellular fibroma-like MP, cellular immature MP, intravascular MP, and malignant MP.⁴

Malignant MP lesions are exceptionally rare, and we are only aware of one case series and an isolated report describing their natural history.^{3,4} Of the five cases presented in the largest series, three were located in the lower extremities, one in the neck, and one in the mediastinum.³

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Around 80% of lesions showed distant metastasis, with the most common sites being liver and brain. Three patients died of their disease within 1 year of presentation, one remained alive at 24 months following diagnosis, and one had no recurrence following wide excision at 18 months. Concerning histopathological findings of a malignant MP include elevated cellularity, a high degree of mitotic activi-

ty, pleomorphism, and necrosis,³ and warrant prompt surgical referral.

Methods and Results

A systematic literature search was conducted using the PubMed, EMBASE, and Google Scholar databases. The search

Table 1 Systematic literature review of cases of hand and wrist myopericytoma

Source	Age	Sex	Site	Depth	Presentation	Tx	Re-currence	FU
Granter et al ¹	66	M	HTE	UK	UK	UK	UK	24 mo
Dray et al ¹³	60	M	Hand (dorsum)	UK	UK	Exc	UK	UK
	30	M	Hand (acral) ^a	D	UK	Exc	UK	UK
Mentzel et al ⁴	55	M	Finger	D/SC	UK	Exc	UK	UK
	57	M	Hand (dorsum)	SC	UK	Exc	UK	UK
	70	M	Hand	D	UK	Exc	UK	UK
	13	F	Finger	D	UK	Exc	UK	UK
	34	M	Hand	D	UK	Exc	UK	UK
	20	F	Hand	SC	UK	Exc	UK	UK
	34	M	Hand	D	UK	Exc	UK	UK
	52	M	Finger	D/SC	UK	Exc	UK	UK
	62	M	Finger	SC	UK	Exc	UK	UK
Sadahira et al ⁸	70	M	Periungual	SC	UK	Obs	N/A	8 mo ^b
Chalmers et al ¹⁷	63	M	SPA	Superficial to flexor tendons	Intermittently painful spherical swelling	Exc	UK	UK
Harreld et al ¹⁸	36	F	SPA	Superficial to palmar digital nerve	Intermittent stabbing pain left palm	Exc	No	24 months
Díaz-Flores et al ¹⁹	76	F	Wrist	IV	UK	UK	UK	UK
Kara et al ¹¹	18	M	SPA	Superficial to palmar digital nerve	Painful mass in thenar region + numbness ulnar thumb	Exc	No	14 mo
Aung et al ⁵	45	F	Wrist (dorsum)	D	Scaly, keratotic papule	Exc	No	UK
Mahaptra et al ²⁰	59	F	Digital artery	IV	Painful, gradually increasing mass	Exc	No	3 mo
Wagner et al ²¹	58	M	HTE	Hypothenar adipose tissue	Wrist pain, numbness/paresthesia	Exc	UK	UK
Orenay et al ⁹	72	M	Wrist	SC		Exc	No	1 mo
Michot et al ²²	31	F	HTE	IV	Painless, gradually increasing in size, resulting in Guyon's canal syndrome	Exc	No	4 y

Abbreviations: D, dermis; Exc, excision; F, female; HTE, hypothenar eminence; IV, intravascular; M, male; N/A, not applicable; Obs, observation; SC, subcutis; SPA, superficial palmar arch; UK, unknown.

^aIdentified as "probably malignant."

^bNo progression at 8 months.

term that was utilized was “myopericytoma” and this yielded only 22 cases of MP presenting in the hand and wrist (►Table 1). Conference abstracts and articles not written in English were excluded. Of the 22 cases identified, 6 lesions were located in the finger, 13 were located in the hand, and 3 were located in the wrist. Of those located in the finger, one directly invaded the radial digital artery. Of the palmar cases, three were determined to involve the superficial palmar arch. Unfortunately, many of the cases documented have an unknown clinical presentation and medical history. Of those cases with available information, all except one lesion presented as a painful nodule. No cases showed evidence of lytic destruction. In two of the cases, patients reported paresthesias and numbness. There were two noted comorbidities (chronic obstructive pulmonary disease⁸ and T-cell lymphoma⁹). The mean age of patients with hand and wrist involvement was 49 years (standard deviation = 19) with a range of 13 to 76 years. There was a strong male predominance (68%). All cases, except for one, where surgery was declined, were managed by excision. There were no documented cases of recurrence.

Case Report

A 33-year-old right-hand dominant male presented to our outpatient clinic with a 2-month history of painless swelling and erythema of the pulp of his left index finger (►Fig. 1). He was otherwise healthy and had no history of trauma. At the time of presentation, he was enrolled in a fine arts program and was an active guitarist. On physical examination, his finger was noted to be firm and erythematous, but not tender or fluctuant. He was neurovascularly intact. His range of motion was within normal limits, with full movement of his flexor digitorum superficialis, flexor digitorum profundus, and extensor tendons. Contralateral hand examination was unremarkable. Plain film investigations showed the extensive bony destruction of the distal phalanx (►Fig. 2), which was suggestive of a primary or secondary neoplastic process. An incisional biopsy was undertaken in the outpatient clinic which showed

Fig. 1 Dorsal (A) and lateral (B) views of the patient's left index finger.

Fig. 2 Anteroposterior (A) and lateral (B) plain film X-ray of the patient's left index finger.

cellular characteristics of MP: oval-to-spindle-shaped cellular architecture with abundant eosinophilic cytoplasm and an indistinct cellular border. Additional characteristics included vascular luminal invasion and minimal atypia. Immunohistochemical analysis showed uniform expression of vimentin and weakly patchy expression of smooth muscle actin.

The patient returned to the clinic for follow-up 4 weeks post incisional biopsy. At this time he was noted to have some improvement in the swelling and erythema of his digit. After extensive discussion of his surgical options, including distal amputation with preservation of the flexor digitorum profundus tendon and excision with tumor shelling, he decided to proceed with the latter. The patient subsequently underwent left index finger tumor resection with local advancement flap closure. The procedure was unremarkable. Final pathology showed the extension of the lesion to the resection margins.

At 81-day follow-up, the patient was pleased with the overall results of the procedure and had resumed his artistic activities (►Figs. 3–5). There was no evidence of tumor recurrence on physical examination. The patient did complain of some residual feeling of lack of support distally with hard pinch, but we reassured him that this would continue to improve over time.

Discussion

Imaging of the hands, wrists, and fingers begins with plain film X-ray in the anteroposterior, lateral, and oblique views. Attention should be paid to the number of lesions present and their location, periosteal reactions, the degree of lytic destruction, and the degree of soft tissue involvement.¹⁰

Fig. 3 Dorsal view of the left hand showing healed tumor excision site.

Fig. 4 Lateral view of the left hand showing healed tumor excision site.

Fig. 5 Superior view of a left hand clenched grip and healed tumor excision site.

There are however limitations to conventional radiographs for tumors of the upper extremities. Other imaging modalities, including ultrasonography and magnetic resonance (MR) imaging, provide useful diagnostic information and

should be considered if available to the clinician. Ultrasound may be particularly useful in the initial evaluation of soft tissue masses, particularly with concomitant tendon or pulley concerns.¹⁰ MR imaging is the superior modality for assessing lytic and soft tissues lesions of the upper extremities. In the case presented, an MR was not obtained given the latency to presentation. In previous studies, MP lesions appear lobulated, well circumscribed, and mass enhancing on MR. T1A- and T2A-weighted images demonstrate low- and high-signal intensity, respectively.¹¹

There are several biopsy modalities for the assessment of tumors presenting in the hand and wrist. These include excisional biopsy, fine needle aspiration (FNA), and core biopsy. The authors selected an excisional biopsy in the aim of improving the patient's digital function as a bridge to definitive treatment. Although excisional biopsies have been shown to be superior to both FNA and core biopsy in the diagnosis of soft tissue lesions, they carry a greater risk of tumor spread and complication.¹²

Myopericytoma is difficult to distinguish from other lytic and soft tissue lesions on the basis of clinical and radiographic evidence alone. From our experience and literature analysis, MP tumors do not have a distinct presentation or any pathognomonic features. Similarly, findings on plain film and MR are not discrete, and many characteristics overlap with those of other clinical entities. Although radiographic evidence is an essential first step in the characterization of primary tumors of the hand and wrist, a biopsy is ultimately required for the diagnosis and management of MP.

MP lesions are a poorly studied clinical entity and are of unknown etiology. To date, only isolated case reports have been described in the primary literature, and thus may be overlooked on differential diagnosis. Furthermore, after our extensive literature review, their clinical presentation appears inherently heterogeneous. Although MP lesions tend to grow slowly and many are benign, physicians should be aware of the possibility of malignancy or progression to malignancy. If the histological analysis shows diffuse mitotic activity, necrosis, and pleomorphism, aggressive treatment is warranted as these lesions carry the potential for morbidity and mortality. Alternatively, MP tumors showing benign histopathological characteristics have little evidence for recurrence, suggesting that they can be managed more conservatively by simple resection or tumor shelling.

Although there is a paucity of evidence, our review of the literature found that, in general, MP lesions are indolent, and develop slowly, unless displaying malignant features (i.e., necrosis, abnormal mitotic activity). There have been very few reports of local recurrence after excision,⁵ and in one of the largest series of patients, there were only two cases of recurrence in 46 tumors at 48 months follow-up, despite incomplete excision in nearly 50% of cases.⁴ Reports of spontaneous regression have also been reported, further demonstrating their indolent nature.^{1,13} In locally aggressive tumors of the hand and wrist, definitive pathology will ultimately determine surgical management, but there is a trend toward tissue conservation in even the most aggressive lesions.^{14,15} Non-malignant lesions of the upper extremity

causing lytic destruction, including tumors and cysts,¹⁶ necessitate clear excision to prevent further invasion. Herein, our patient requested preservation of the distal pharynx, and despite best efforts, clear resection margins were not attained. Given the patient's unique occupation, the location of his tumor, the lesions' benign pathological characteristics, and the literature available to date, tumor shelling was considered, and may be a reasonable primary treatment option in select cases.

This case report and systematic literature review is limited by its number of patients and lack of long-term follow-up, but guides the evaluation, diagnosis, and management of MP when presenting in the hand and wrist. Additionally, the case presented herein highlights the challenges faced when determining the treatment course in an otherwise healthy, active individual with unique functional demands.

Note

A written informed consent was obtained from all patients for being included in this study.

Funding

None.

Conflict of Interest

None.

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