Myopericytoma of the distal forearm: a case report

Guang Yang · Evan P. McGlinn · Kevin C. Chung

Published online: 16 May 2014
© American Association for Hand Surgery 2014

Abstract Myopericytoma is a rare type of soft tissue tumor with perivascular myoid differentiation. Although the pathology characteristics of myopericytomas are well described in literature, the clinical characteristics of these tumors have received less attention. We report on a 44-year-old female who developed a myopericytoma in her right distal forearm. The patient presented with a painless solid mass that had been slowly increasing in size for approximately 5 years. Unlike typical myopericytoma in the extremity growing as a subcutaneous nodule, the tumor enveloped the distal ulna. This case suggests a different growth pattern for myopericytoma. Myopericytoma is a rare soft tissue tumor originating from perivascular myoid cells, which has only been recognized as a distinct condition in the past 15 years (Granter et al. *Am J Surg Pathol.* 22 (5):513–25, 1998). Most of these tumors present as a painless, slow-growing subcutaneous nodule. They have typically been described by pathologists who concentrate on the pathological features rather than the clinical characteristics of these tumors (Granter et al. *Am J Surg Pathol.* 22 (5):513–25, 1998; Dray et al. *J Clin Pathol.* 59 (1):67–73, 2006; Mentzel et al. *Am J Surg Pathol.* 30 (1):104–113 2006). We report a case of myopericytoma with an unusual growth pattern involving the distal forearm to highlight the clinico-pathologic features of this tumor.

Case Report

A 44-year-old woman presented with a slow-growing mass that was noted at the ulnar aspect of the right distal forearm. The tumor had been present for approximately 5 years. She had no pain in the area. The patient had a history of an intraneural glomus tumor with unknown malignant potential on the median nerve in the same side of the upper arm dating back to 1993. A subtotal resection of the glomus tumor and postoperative radiotherapy were performed in 2001. She had been disease free for the past 12 years. Our physical examination showed a round 2-cm mass over the ulnar aspect of the wrist. The mass was firm, nonpulsatile, and without a thrill. Her ulnar nerve function was normal. She had partial loss of the function of the median nerve owing to the effects of the prior glomus tumor resection and radiation to the median nerve. There was no lymphadenopathy.

Ultrasound showed a complex, hypoechoic, well-circumscribed, solid mass. The size of the mass was $2.1 \times 2.8 \times 1.5$ cm, with increased vascularity with color Doppler imaging. Magnetic resonance imaging (MRI) with gadolinium contrast revealed a heterogeneously enhancing mass extending around the distal ulna. The mass was in direct contact with the volar aspect of the extensor carpi ulnaris tendon without erosion of the bone (Fig. 1). Furthermore, the patient had a chest radiograph to rule out metastasis. There was no evidence of metastasis before surgery.

Surgical biopsy was planned. We made a longitudinal incision on the ulnar side of the distal forearm, with the patient under general anesthesia. The tumor was a solid soft tissue mass with yellow, soft appearance. We excised a sufficient piece to send for permanent pathology. The permanent pathology identified a benign tumor, which was diagnosed as myopericytoma. The complete excision of the tumor was undertaken after 2 months. The incision was carried out along the former incision. The ulnar neurovascular bundles as well

G. Yang
Department of Hand Surgery, China-Japan Union Hospital of Jilin University, Changchun, China

E. P. McGlinn · K. C. Chung (✉)
Section of Plastic Surgery, Department of Surgery, The University of Michigan Health System, 2130 Taubman Center, SPC 5340, 1500 E. Medical Center Drive, Ann Arbor, MI 48109-5340, USA
e-mail: kecchung@med.umich.edu

Fig. 1 MRI with gadolinium contrast shows a heterogeneously enhancing mass (*red arrow*) surrounding the distal ulnar diaphysis (*yellow arrow*). The mass was in direct contact with the volar aspect of the extensor carpi ulnaris tendon (*blue arrow*) without erosion of the bone and tendons. **a** Sagittal section at the distal forearm, **b** coronal section at the wrist and distal forearm

as the dorsal sensory branch of the ulnar nerve were identified and preserved. We found the tumor wrapped around the interosseous membrane primarily attached to the distal ulna, but there was a plane of demarcation between the tumor and the surrounding soft tissues. The tumor was sharply excised by stripping the periosteum off the ulna and also removing a portion of the interosseous membrane. The entire mass measured about 4.0×2.0 cm and was sent for final pathology examination (Fig. 2). Pathology slides demonstrated elongated and ectatic blood vessels surrounded by spindled cells. At

Fig. 2 Intraoperative image of the patient. **a** The tumor had a yellow, soft appearance after the ulnar neurovascular bundles were retracted. **b** The ulna and dorsal sensory branch of the ulnar nerve were intact after the entire mass was excised with its periosteum off the ulna and also a portion of the interosseous membrane. **c** The excised tumor measured 4.0×2.0 cm

high magnification, the tumor cells surrounding smaller blood vessels are “spiraling off” into the stroma (Fig. 3). The cells themselves were bland and spindled. The cell borders were not readily apparent, and they had eosinophilic cytoplasm. There was no evidence of malignancy.

Her postoperative course was uneventful. At 5-month follow-up, the patient had normal sensation in the ulnar nerve distribution, and intrinsic muscles were of normal bulk and strength. There was no sign of local recurrence and metastasis.

Fig. 3 Photomicrograph of resected specimen (hematoxylin-eosin stain). Tumor cells surrounding smaller blood vessels “spiraling off” into the stroma (magnification $\times 400$)

Discussion

The term myopericytoma was proposed by Granter in 1998 to describe rare tumors of the extremities in seven cases [7]. Myopericytomas originate from myopericytes that have features of pericytes and vascular smooth muscle cells [4]. In 2002, myopericytoma was classified as an independent-type pericytic (perivascular) tumor neoplasm by the World Health Organization [6].

Myopericytoma typically arises in adults but may occur at any age. According to the largest case report ($n=54$), the most common site was the lower extremity with 26 cases occurring at this site, followed by 16 cases in the upper extremities including digits, hands, and arms [13]. The head, neck, and trunk were also affected [13]. The vast majority of lesions arise in the skin and subcutaneous soft tissues, and myopericytomas occurring in the oral cavity [2], thoracic spine [1], lung [16], and kidney [3] have also been reported. Our case presented a single, painless, well-circumscribed, and slow-growing solid mass in the upper extremity. These symptoms are the typical clinical features of myopericytoma (Table 1). The unusual intravascular myopericytomas in extremities usually manifest as painful nodules [8, 11, 18]. In a few rare cases, the lesions presented multicentric and multiple nodules [12, 13, 16]. Although malignant myopericytomas are rare, they have been reported [12, 13]. McMenamin and Fletcher reported three cases of malignant myopericytomas of the extremities, and two of them presented as large, painful deep masses with infiltrative growth and metastasis [12]. Although a few reports in the literature support the possible association between myopericytoma and trauma [9] or chronic scars [15], the pathogenesis of myopericytoma is unknown.

The tumor in this case encased the distal ulna diaphysis with approximately 270° of surface contact with the cortex and no invasion of the tendons and bones. Unlike the typical

Table 1 Clinical features in reported cases of myopericytomas of the extremities

Author (published year)	Number of cases	Age (years)	Sex (M:F)	Site	Presenting symptoms	Size (cm)	Treatment	Outcome
Granter SR (1998)	7	10–66	5:2	Hand, arm, leg, and ankle	Painless nodule in subcutaneous and superficial soft tissue	0.6–1.7	Excision	One case recurred twice in 3 years Lost
McMenamin ME (2002)	1	54	M	Within a vein of the posterior thigh	Slowly growing painful subcutaneous nodule	1.5	Complete excision with an overlying ellipse of skin	
McMenamin ME (2002)	3	19–81	2:1	Thigh, heel of foot, and upper arm	Two cases with a painful mass and one with a superficial painless mass	1.5–13	Wide local excision and radiation; below knee amputation; local excision followed by wide excision	All were malignant, and two cases died of metastases within 1 year
Dray MS (2005)	7	13–71	4:3	Hand, knee, ankle, and foot	Two cases with painful lesions and five with mass lesions	1.1–3.0	Incisional biopsy, excision, followed by either complete excision or close observation	All were free of disease
Mentzel T (2006)	42	13–87	27:15	Upper and lower extremities	Neoplasms of skin and soft tissues	Did not report	Complete or marginal excision	One malignant and one intravascular case recurred
Woollard AC (2007)	1	63	M	Within the superficial palmar arch	Spherical swelling in mid-palm with pain	1.0	Excision	Did not report outcome
Pack JO (2011)	1	45	F	Lower leg	Slow-growing, painless subcutaneous firm nodule	0.9 $\times$ 0.7	Excision	Did not report outcome
Ko JY (2011)	1	67	M	Within a vein of the lateral thigh	Painful, slow-growing, skin-colored hard subcutaneous nodule	3.0 $\times$ 3.5	Excision	Did not report outcome

myopericytoma in the extremity that presents as a subcutaneous nodule, this unique growth pattern has not been reported before.

Although the presenting symptoms in this case were typical, they were not specific to myopericytoma. Rho et al. reported ultrasound-guided core-needle biopsy was used for diagnosis in a case in the neck [14]. Considering the location of the mass, we selected incisional biopsy to avoid potential nerve and vessel injury. The pathological differential diagnosis of myopericytoma includes myofibroma, angioleiomyoma, and glomus tumor because of histologic overlap [13]. On microscopic examination, myopericytomas contain elongated and ectatic blood vessels concentrically surrounded by spindle myoid tumor cells with eosinophilic cytoplasm. The presence of a zonation/biphasic appearance is most suggestive of myofibroma rather than myopericytoma [5]. Most angioleiomyomas are comprised of thick-walled vessels and present a desmin-positive immunohistochemical profile [10, 13]. In contrast, most myopericytomas have thin-walled vessels and are negative for desmin [10, 13]. Glomangioma cells have the perivascular arrangement but lack the spindle cells [13]. The other immunohistochemical features of myopericytomas are positive for vimentin, smooth muscle actin, muscle-specific actin, and often for h-caldesmon, whereas S-100 protein, P-53 protein, and CD99 are usually negative [13, 14, 17].

Most myopericytomas are benign tumors and are treated by complete excision with clear margin. Recurrences are caused by incomplete tumor excision [13]. Although we cannot make a definitive statement since the follow-up in our patient was only 5 months, we believe surgical excision is sufficient for treatment after the pathology diagnosis of benign myopericytoma is made.

Conflict of interest Guang Yang does not have a conflict of interest.

Evan P. McGlinn does not have a conflict of interest.

Kevin C. Chung does not have a conflict of interest.

Financial Disclosure Kevin C. Chung declares that the research reported in this publication was supported by the National Institute of Arthritis and Musculoskeletal and Skin Diseases of the National Institutes of Health under Award Number K24 AR053120. The content is solely the responsibility of the authors and does not necessarily represent the official views of the National Institutes of Health.

Guang Yang has no financial disclosure.

Evan P. McGlinn has no financial disclosure.

Statement of Human and Animal Rights All the authors declare that 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 (5).

Statement of Informed Consent Informed consent was obtained from all patients for being included in the study. All the authors declare that this manuscript contains no identifying information.

References

1. Agrawal N, Nag K. Myopericytoma of the thoracic spine: a case report and review of literature. *Spine J*. 2013;13(11):e23–7.
2. Datta V, Rawal YB, Mincer HH, et al. Myopericytoma of the oral cavity. *Head Neck*. 2007;29(6):605–8.
3. Dhingra S, Ayala A, Chai H, et al. Renal myopericytoma: case report and review of literature. *Arch Pathol Lab Med*. 2012;136(5):563–6.
4. Dictor M, Elner A, Andersson T, et al. Myofibromatosis-like hemangiopericytoma metastasizing as differentiated vascular smooth-muscle and myosarcoma. Myopericytes as a subset of “myofibroblasts”. *Am J Surg Pathol*. 1992;16(12):1239–47.
5. Dray MS, McCarthy SW, Palmer AA, et al. Myopericytoma: a unifying term for a spectrum of tumours that show overlapping features with myofibroma. A review of 14 cases. *J Clin Pathol*. 2006;59(1):67–73.
6. Folpe AL, McMenamin ME. Pericytic (perivascular) tumors. In: Fletcher CDM, Unni KK, Mertens F, editors. WHO classification of tumours, pathology and genetics of tumours of soft tissue and bone. Lyon: IARC Press; 2002. p. 135–9.
7. Granter SR, Badizadegan K, Fletcher CD. Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: a spectrum of tumors showing perivascular myoid differentiation. *Am J Surg Pathol*. 1998;22(5):513–25.
8. Ko JY, Choi WJ, Kang HS, et al. Intravascular myopericytoma: an interesting case of a long-standing large, painful subcutaneous tumor. *Pathol Int*. 2011;61(3):161–4.
9. Laga AC, Tajirian AL, Islam MN, et al. Myopericytoma: report of two cases associated with trauma. *J Cutan Pathol*. 2008;35(9):866–70.
10. Matsuyama A, Hisaoka M, Hashimoto H. Angioleiomyoma: a clinicopathologic and immunohistochemical reappraisal with special reference to the correlation with myopericytoma. *Hum Pathol*. 2007;38(4):645–51.
11. McMenamin ME, Calonje E. Intravascular myopericytoma. *J Cutan Pathol*. 2002;29(9):557–61.
12. McMenamin ME, Fletcher CD. Malignant myopericytoma: expanding the spectrum of tumours with myopericytic differentiation. *Histopathology*. 2002;41(5):450–6.
13. Mentzel T, Dei Tos AP, Sapi Z, et al. Myopericytoma of skin and soft tissues: clinicopathologic and immunohistochemical study of 54 cases. *Am J Surg Pathol*. 2006;30(1):104–13.
14. Rho BH, Lee SK, Kwon SY. Myopericytoma of the neck: sonographic appearance and sonographically guided needle biopsy. *J Clin Ultrasound*. 2011;39(8):469–72.
15. Scott RS, Blank KL, Proffer LH, et al. Perivascular myoma of myopericytoma and myofibromatosis-type arising in a chronic scar. *J Cutan Pathol*. 2006;33(3):231–5.
16. Song XL, Sun XY, Zhang GL, et al. Myopericytoma presenting as multiple pulmonary nodules. *Intern Med*. 2012;51(6):639–42.
17. Terada T. Minute myopericytoma of the neck: a case report with literature review and differential diagnosis. *Pathol Oncol Res*. 2010;16(4):613–6.
18. Woollard AC, Southgate C, Blair JW. Intravascular myopericytoma of the superficial palmar arch. *J Hand Surg Eur*. 2007;32(4):475–6.