Painful Forearm Mass in a 75-year-old Man

Scott J. Schoenleber MD, Andrew E. Rosenberg MD,
H. Thomas Temple MD

Received: 23 August 2013 / Accepted: 22 November 2013 / Published online: 4 December 2013
© The Association of Bone and Joint Surgeons® 2013

History and Physical Examination

A 75-year-old right-hand-dominant man presented with a 3-year history of a painful left forearm mass. The pain was nonradiating and throbbing in nature, present at night, and the mass was exquisitely tender to palpation. The mass did not change in size throughout the day or with exercise. There was no cold intolerance, antecedent trauma, or constitutional symptoms. The medical history was otherwise unremarkable and there was no family history of neoplasia.

Approximately 1.5 years before presentation, the patient underwent an attempted excisional biopsy of the lesion. The pathology at that time was not available for review, but the patient was informed that it was nondiagnostic. Subsequently, the patient had no improvement of symptoms. In fact, in the months after the biopsy the mass increased in

size and became more painful. He presented for further evaluation and treatment.

On examination, there was mild bluish discoloration of the skin overlying the proximal dorsomedial forearm. There was a well-healed transverse surgical incision distal to the point of maximal tenderness. There was exquisite tenderness to palpation of a 1-cm, mobile, subcutaneous mass. There was a negative percussion sign, negative transillumination test, and no lymphadenopathy or other masses. The patient had full elbow ROM and a normal neurovascular examination.

The patient initially presented with recent MR images (Figs. 1, 2). Radiographs of the affected elbow were normal. An office ultrasound also was performed to localize and to better characterize the lesion (Fig. 3).

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

Each author certifies that he or she, or a member of his or her immediate family, has no commercial associations (eg, consultancies, stock ownership, equity interest, patent/licensing arrangements, etc) that might pose a conflict of interest in connection with the submitted article.

All ICMJE Conflict of Interest Forms for authors and *Clinical Orthopaedics and Related Research* editors and board members are on file with the publication and can be viewed on request. 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.

S. J. Schoenleber, H. T. Temple (✉)
Division of Musculoskeletal Oncology, Department of
Orthopaedic Surgery, University of Miami, 1400 NW 12th
Avenue, Room 4036, Miami, FL 33136, USA
e-mail: htemple@med.miami.edu

A. E. Rosenberg
Department of Pathology, University of Miami, Miami, FL, USA

Imaging Interpretation

Ultrasound showed a hypoechoic, homogeneous, well-circumscribed mass in the deep subcutaneous tissues adjacent to the fascia of the proximal dorsomedial forearm (Fig. 3). Doppler imaging confirmed the presence of vascularity within the mass. Magnetic resonance imaging demonstrated a homogeneous, well-circumscribed, 10 × 6 mm mass with decreased signal intensity on T1 (Fig. 1) and increased intensity on T2 fat saturation imaging located in the deep subcutaneous tissues. T1 weighted fat saturation imaging demonstrated increased signal intensity, which uniformly enhanced with contrast (Fig. 2). There was no surrounding edema and the mass appeared to be well encapsulated.

Fig. 1 A coronal T1-weighted MR image reveals a low-signal-intensity 10-mm mass in the deep dorsomedial subcutaneous tissues of the left forearm (arrow).

Differential Diagnosis

Extradigital glomus tumor (solid type, glomangioma, glomangiomyoma)

Angiolipoma

Vascular malformation (hemangioma)

Angioleiomyoma

Epithelioid hemangioma

Schwannoma

The lesion was excised, and frozen sections were evaluated histologically (Figs. 3, 4).

Based on the history, physical examination, imaging studies, and histopathology, what is the diagnosis and how should the patient be treated?

Histology Interpretation

Gross histologic examination shows a well-circumscribed, pink-gray nodule measuring $7 \times 5 \times 3$ mm. Low-power histologic examination shows a well-circumscribed mass composed of uniform round cells with central nuclei, eosinophilic cytoplasm with well-formed cytoplasmic membranes, and a stroma that contains numerous dilated thin-walled vessels (Fig. 4). Some of the tumor cells cuff the blood vessel walls. There is no significant atypia or mitotic activity (Fig. 5).

Fig. 2A–B Axial T1-weighted fat saturation images (A) before and (B) postgadolinium show a homogeneous deep subcutaneous mass with increased signal intensity and uniform enhancement with contrast (arrow), with an overlying radiographic marker (arrowhead) in the region of pain.

Diagnosis

Extradigital glomangioma.

Discussion and Treatment

In this patient, the pertinent findings supporting the diagnosis of an extradigital glomangioma include the presence of an exquisitely painful mass with overlying skin discoloration. MRI findings, while nonspecific, were more typical of those seen in the solid subtype of glomus tumor: a small, subcutaneous, encapsulated mass with low intensity on T1 and high intensity on T2 sequences. Histologic examination confirmed the diagnosis of glomangioma, with characteristic findings of monomorphic sheets of

Fig. 3 An ultrasound image of the proximal forearm shows a hypoechoic, well-circumscribed mass (A) located in the subcutaneous tissues (white double arrow) adjacent to the fascia (black arrows). The dimensions of the mass as recorded at the time of testing are indicated by the faint gray numbers 1 and 2.

Fig. 4 A low-power microscopic view shows sheets of uniform round cells with central nuclei, eosinophilic cytoplasm, and a stroma containing numerous dilated thin-walled vessels (Stain, hematoxylin and eosin; original magnification, $\times 100$).

moderately sized cells with uniform nuclei and eosinophilic cytoplasm, with a stroma containing numerous dilated, thin-walled vessels. Glomus tumors have been reported infrequently in superficial, deep, and intravascular locations of the forearm, with variable symptoms [1, 6, 11]. Glomangioma also has been reported in the forearm, resulting in compression neuritis [5]. This case was unique in that the imaging and clinical features were more suggestive of a solid-type glomus tumor. It illustrates that

Fig. 5 A high-power microscopic view shows uniform cells, some of which cuff the blood vessels, without atypia or mitotic activity (Stain, hematoxylin and eosin; original magnification, $\times 200$).

uncommon neoplasms may present with atypical characteristics, reinforcing the need for tissue diagnosis.

The differential diagnosis was broad, and based on the history and physical and the imaging studies, primarily focused on benign vascular tumors. Angiolipoma may present as a soft, mobile, subcutaneous mass with pain on palpation. These most commonly occur in the forearm, but are rare in patients older than 50 years. Vascular malformation can be a cause of an aching subcutaneous mass. Patients often will report activity-related discomfort and a mass that fluctuates in size, which may improve with compression wraps and antiinflammatory medications. Radiographs may show phleboliths, which are small, rounded mineralizations. Angioleiomyoma, also called angiomyoma or vascular leiomyoma, was considered as these tumors often will present in middle age as painful solitary lesions in the subcutaneous tissues of the extremities [17]. Upper extremity involvement may be observed in approximately 22% [4]. Occasionally, angioleiomyomata may mineralize, especially in acral locations [8]. These benign tumors are often painful, usually occur in the extremities, and are slow growing [14]. A less likely possibility was epithelioid hemangioma, which may present clinically as a superficial or deep soft tissue mass. Typically, these occur in middle age. The majority occur in the head and neck and they may be associated with overlying skin changes such as plaques or excoriations. Lymphadenopathy may be present [18]. Schwannomas are painful, usually solitary soft tissue masses that are slowly growing. They typically are associated with radiating pain on percussion, and often will exhibit a “string sign” and, occasionally, a “bull’s eye sign” on MRI. Multiple potential diagnoses were considered at the time of excision.

Extradigital glomus tumors, including glomangioma, often are unexpected and difficult to diagnose. Unlike

digital glomus tumors, which often present with a triad of pain, tenderness to palpation, and cold intolerance, extradigital glomus tumors have a more variable clinical presentation and rarely present with cold hypersensitivity [15]. These benign tumors have a male-to-female predominance greater than 4 to 1 and can present at any time during adulthood [7, 15]. Skin discoloration, as seen in our patient, is a common finding [7, 15]. Glomangioma may present with similar clinical features, but in contrast to the solid subtype, accounts for only approximately 20% of glomus tumor subtypes [15], usually presents in childhood or early adulthood, and is less likely to be painful [19].

Histopathologically, glomus tumors arise from the neuromyoarterial glomus body, which plays a role in the regulation of blood flow to the skin. There are three subtypes of glomus tumor, based on the pattern of differentiation and the degree of vascularity. These include the solid type, which is the most frequent variant and is known as a “glomus tumor.” These are characterized by sheets of eosinophilic, polygonal cells surrounding capillaries [15]. Glomangioma, as seen in our patient, is a variant with cavernous vascular spaces that are cuffed by glomus cells, which also may grow in sheets. The third subtype, glomangiomyoma, exhibits glomus cells associated with smooth muscle differentiation [7, 15].

Although the natural history of extradigital glomus tumors is typically that of a benign process, patients often are misdiagnosed for years because of the relative infrequency and atypical presentation of these lesions [2, 7, 10, 15, 20]. Patients can experience excruciating pain and difficulty with sleep. It is not uncommon for symptoms to be present for years before diagnosis is made [2, 7, 15].

Nonspecific radiographic findings may contribute to the diagnostic challenge. Radiographs, as in our patient, are frequently nondiagnostic [15]. Ultrasound is useful to differentiate solid tumors from cystic masses. Ultrasonographically, glomus tumors tend to be ovoid, richly vascular, well circumscribed, and display mixed- or hypoechoogenicity [12]. Glomangioma, in general, tends to be more poorly circumscribed than the solid subtype. MRI remains the most reliable imaging modality, with a 100% success rate in identifying the mass in one study [15]. Postcontrast enhancement is typical [9]. However, findings of low intensity on T1 and increased intensity on T2 are nonspecific and are observed with many soft tissue neoplasms. The imaging characteristics in this case were somewhat unusual for a glomangioma, given the relatively well-circumscribed appearance on imaging, which reinforces the need to correlate the imaging findings with the histopathologic findings.

A glomus tumor, regardless of subtype, is treated by marginal excision when symptomatic. This generally is curative in 90% or more of the patients [7, 15]. Others

report slightly higher rates of local recurrence, ranging from 13% to more than 30%, which may be attributable to either incomplete resection or true recurrence [2, 16]. Although there have been isolated reports of glomangiomas with aggressive features, including a report of a locally aggressive glomangioma, this is rare [3]. Glomangiosarcoma, the term for a malignant glomus tumor, is extremely rare [3, 13].

Our patient was treated with extracapsular marginal resection. We elected not to perform a wide local excision as this was not felt to represent a true recurrence. Rather, based on the original reported pathology and the key physical examination finding of a transverse incision several centimeters distal to the mass, we felt that the initial treating surgeon most likely removed nonpathologic subcutaneous tissue at the index procedure. Intraoperative ultrasound was used for precise localization given the history of failed excision. Although transverse incisions should be avoided for tumor excision because they are not extensile and can contaminate multiple tissue planes, the benign appearance on imaging in this case allowed us to approach this mass through a new longitudinal incision without any attempt at excision of the previous biopsy tract. Deep subcutaneous dissection revealed a well-encapsulated mass with a blue-purple hue. Frozen section was obtained and was suggestive of glomangioma. The diagnosis of extradigital glomangioma was confirmed on final pathologic examination based on the cellular appearance and the degree of vascularity. At the first postoperative followup the patient was doing well and had complete resolution of his symptoms. This case reinforces the fact that misdiagnosis of extradigital glomus tumors of any subtype is common, attributable in part to atypical presentations and nonspecific imaging characteristics. Meticulous surgical technique, including intraoperative imaging as necessary, is required for tumors located in the deep subcutaneous tissues.

References

1. Beham A, Fletcher CDM. Intravascular glomus tumour: a previously undescribed phenomenon. *Virchows Archiv A Pathol Anat Histopathol.* 1991;418:175–177.
2. Fazwi R, Chandran PA, Ahmad TS. Glomus tumour: a retrospective review of 15 years experience in a single institution. *Malaysian Orthop J.* 2011;5:8–12.
3. Gould EW, Manivel JC, Albores-Saavedra J, Monforte H. Locally infiltrative glomus tumors and glomangiosarcomas: a clinical, ultrastructural, and immunohistochemical study. *Cancer.* 1990;65:310–318.
4. Hachisuga T, Hashimoto H, Enjoji M. Angioleiomyoma: a clinicopathologic reappraisal of 562 cases. *Cancer.* 1984;54:126–130.
5. Jiga LP, Rata A, Ignatiadis I, Geishauser M, Ionac M. Atypical venous glomangioma causing chronic compression of the radial

- sensory nerve in the forearm: a case report and review of the literature. *Microsurgery*. 2012;32:231–234.
6. Kogure H, Shirakura K, Arisawa N, Aramaki T, Shimegi A. 3 deep-seated glomus tumors in the lower arm. *Acta Orthop Scand*. 1996;67:194–195.
 7. Lee DW, Yang JH, Chang S, Won CH, Lee MW, Choi JH, Moon KC. Clinical and pathological characteristics of extradigital and digital glomus tumours: a retrospective comparative study. *J Europ Acad Dermatol Venereol*. 2011;25:1392–1397.
 8. Maheshwari AV, Temple HT, Murocacho CA. Calcified angio-myomas of the foot: a case report. *Foot Ankle Int*. 2008;29:449–455.
 9. Matloub HS, Muoneke VN, Prevel CD, Sanger JR, Yousif NJ. Glomus tumor imaging: use of MRI for localization of occult lesions. *J Hand Surg Am*. 1992;17:472–475.
 10. Myers RS, Lo AK, Pawel BR. The glomangioma in the differential diagnosis of vascular malformations. *Ann Plast Surg*. 2006;57:443–446.
 11. Nigam JS, Misra V, Singh A, Karuna V, Chauhan S. A glomus tumour arising from the flexor aspect of the forearm: a case report with review of the literature. *J Clin Diagn Res*. 2012;6:1559–1561.
 12. Park HJ, Kim SS, Lee SO, Choi YJ, Chung EC, Rho MH. Sonographic appearances of soft tissue angioleiomyomas: differences from other circumscribed soft tissue hypervascular tumors. *J Ultrasound Med*. 2012;31:1589–1595.
 13. Park JH, Oh SH, Yang MH, Kim NI. Glomangiosarcoma of the hand: a case report and review of the literature. *J Dermatol*. 2003;30:827–833.
 14. Ramesh P, Annapureddy SR, Khan F, Sutaria PD. Angioleiomyoma: a clinical, pathological and radiological review. *Int J Clin Pract*. 2004;58:587–591.
 15. Schiefer TK, Parker WL, Anakwenze OA, Amadio PC, Inwards CY, Spinner RJ. Extradigital glomus tumors: a 20-year experience. *Mayo Clin Proc*. 2006;81:1337–1344.
 16. Strahan J, Bailie HW. Glomus tumour: a review of 15 clinical cases. *Br J Surg*. 1972;59:91–93.
 17. Weiss SW, Goldblum JR, Folpe AL. Benign tumors of smooth muscle. *Enzinger and Weiss's Soft Tissue Tumors*. 5th ed. St Louis, MO: Mosby; 2008:517–544.
 18. Weiss SW, Goldblum JR, Folpe AL. Benign tumors and tumor-like lesions of blood vessels. *Enzinger and Weiss's Soft Tissue Tumors*. 5th ed. St Louis, MO: Mosby; 2008:644–650.
 19. Weiss SW, Goldblum JR, Folpe AL. Perivascular tumors. *Enzinger and Weiss's Soft Tissue Tumors*. 5th ed. St Louis, MO: Mosby; 2008:751–768.
 20. White CP, Jewer DD. Atypical presentation of a glomus tumour: a case report. *Can J Plast Surg*. 2006;14:237–238.