Simultaneous Solitary Glomus Tumors in Nonadjacent Digits

Todd R. Rimington, MD, and Charles S. Lefton, MD

The glomus tumor is a rare tumor that arises from the glomus apparatus of the dermal retinacular layer of the skin.[1] This tumor has solitary and multiple forms. Solitary tumors are commonly found in the distal phalanx in a paraungual or subungual location. They present with a classic triad of paroxysmal pain, pinpoint tenderness, and cold hypersensitivity.[1] Multiple tumors are extremely rare, are inherited in an autosomal-dominant pattern, and can have involvement outside the hand. Multiple tumors are commonly asymptomatic, but the multiple form can have mixed symptomatic and asymptomatic tumors. Histopathologic characteristics can be used to differentiate the solitary and multiple forms.

To our knowledge, simultaneous solitary glomus tumors in nonadjacent digits have not been reported in the literature. In this article, we describe the case of a woman in her early 40s with clinical, radiographic, and histologic findings for simultaneous solitary glomus tumors in nonadjacent digits of the hand. The authors obtained the patient's written informed consent for print and electronic publication of her case report.

Case Report

A woman in her early 40s presented to our general orthopedic clinic with painful masses on her left index and left ring fingers. The mass on the left index finger had been present for 20 years and had slightly increased in size. The mass on the left ring finger had been present for 5 years and had remained stable in size. The woman presented to our clinic because her paroxysmal pain had become more severe. She had no significant past medical or surgical history. She had no family history of glomus tumor. On examination, she had an 8×5-mm oval blue mass at the ulnar paraungual region of the distal left index finger. The left ring finger had a smaller, 4×2-mm blue mass at the radial paraungual region. At both masses, there was extreme pinpoint tenderness but minimal cold intolerance.

Plain radiographs of the left hand showed the index soft-tissue mass with minimal thinning of the shaft of the distal phalanx. Contrast-enhanced magnetic resonance imaging (MRI) revealed the index and ring finger masses (Figure 1). The masses were bright on T_2-weighted images and isotense to muscle on T_1-weighted images with vigorous enhancement on postcontrast images. No extension into the bone or flexor tendon was observed.

"The treatment for a symptomatic glomus tumor is marginal excision. When the tumor is excised with negative margins, this treatment is curative."

The patient elected surgical excision of both masses. They were excised through a transungual approach by removing the entire nail and making a longitudinal incision through the nail bed. Both tumors were encapsulated. The index finger mass measured 1.4×0.5×0.4 cm, and the ring finger mass measured 0.6×0.3×0.2 cm (Figure 2). Histology confirmed the diagnosis of solid glomus tumor. The tumor was encapsulated with compact cords of round cells with

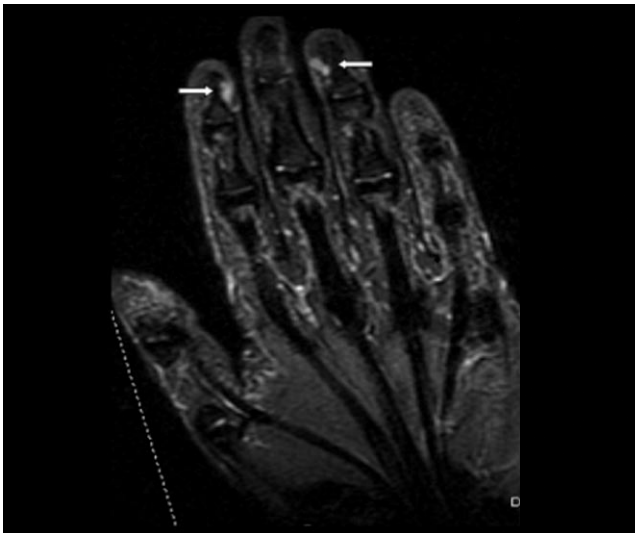

Figure 1. Contrast-enhanced T_2-weighted magnetic resonance imaging of left hand shows bright enhancement of index and ring finger masses.

Dr. Rimington is Resident, Department of Orthopaedic Surgery, Georgetown University Hospital, Washington, DC.
Dr. Lefton is Senior Partner, Commonwealth Orthopaedics, Arlington, Virginia, and Orthopaedic Surgeon, Virginia Hospital Center, Arlington, Virginia.

Address correspondence to: Todd R. Rimington, MD, Department of Orthopaedic Surgery, Georgetown University Hospital, 3800 Reservoir Rd, Ground Floor PHC, Washington, DC 20007 (tel, 202-444-8766; fax, 202-444-8767; e-mail, toddrimington@md.aaos.org).

Am J Orthop. 2009;38(5):E82-E84.

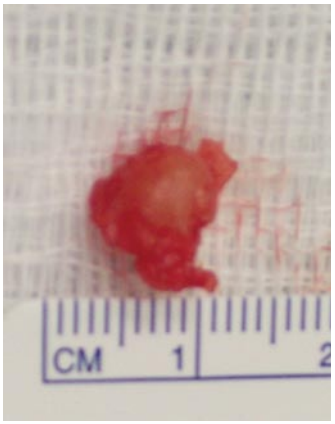

Figure 2. Gross specimen of left index finger glomus tumor shows solitary, simple encapsulated mass measuring 0.6×0.3×0.2 cm.

rounded nuclei without atypia and relatively few vessels (Figure 3). The patient returned to the clinic for a final follow-up 6 months after surgery. By then, her incisions had healed, her fingernails had returned without deformity, and her preoperative symptoms had resolved. There were no clinical signs of recurrence.

Discussion

Glomus tumors arise from the glomus body in the dermal layer of the skin. The glomus body is a myoarterial apparatus that regulates the temperature of the skin. These tumors are rare. They make up less that 2% of all primary soft-tissue tumors and approximately 4% of all tumors in the hand.[1-5] Seventy-five percent of these tumors occur in the hand; a majority, 75% to 90%, occur in the classic subungual location.[1] Glomus tumors can also occur throughout the musculoskeletal, respiratory, urologic, and gastrointestinal systems.[6] The exact etiology of the pain is unknown. One hypothesis is that vasodilation of the Sucquet-Hoyer canal can cause pain directly or indirectly as it compresses the local sensory nerve plexus.[1]

Diagnosis

The typical glomus tumor occurs as a solitary lesion in the hand. The tumor presents with the classic triad of pain, point tenderness, and cold insensitivity. Glomus tumors are more common in women, with a mean age of 30 to 40 years at presentation.[1] Clinically, these tumors are small, less than 1 cm in size, blue, and extremely painful to palpation.

Clinical Diagnostic Testing. Clinical tests that aid in the diagnosis of a glomus tumor include the Love test, the Hildreth test, the transillumination test, and the cold insensitivity test. The Love test is a pain localization test in which a pinhead is used to test the most painful area.[1] The Hildreth test involves modulating the pain produced by the Love test by exsanguinating the digit. The Hildreth test is positive when there is reduction in pinpoint tenderness on exsanguination and ischemia of the digit.[7] The transillumination test reveals an opaque, reddish-bluish mass and allows differentiation from any other masses in the digit.[8] The cold immersion test involves applying an ice cube or ethyl alcohol spray to the mass.[7] The pain associated with a glomus tumor increases with cold application. This test has been found to be highly sensitive and specific.[9]

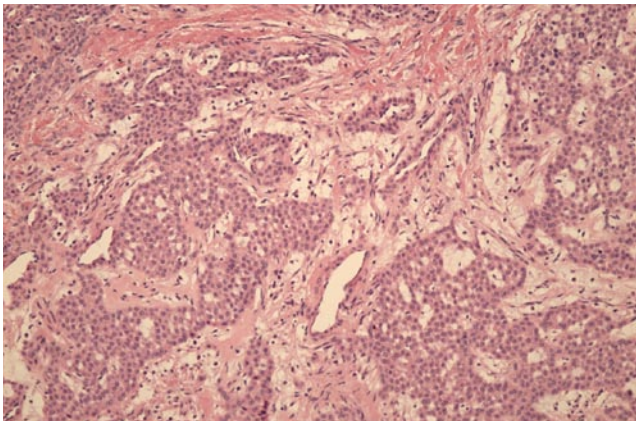

Figure 3. Histology from left index finger glomus tumor shows compact cords of round cells with rounded nuclei without atypia and relatively few vessels.

Imaging Options. Radiographic analysis of glomus tumors includes plain radiography, MRI, magnetic resonance angiography (MRA), high-resolution ultrasonography, and contrast computed tomography (CT). Plain radiographs may show cortical thinning and erosion of the distal phalanx with large tumors. However, positive radiographs have been seen in only 40% of cases.[10] With high-resolution ultrasonography, the tumors appear hypoechoic and have prominent vascularity. However, ultrasound evaluation is limited because of ultrasonographers' lack of experience with glomus tumors and the artifact created by the nail plate.[11] Contrast CT is more commonly used in extradigital locations; the glomus tumors have strong homogenous enhancement in the arterial phase and prolonged enhancement in the delayed phase. MRI is probably the most widely used radiographic study after plain radiography. Glomus tumors have low signal intensity on T_1-weighted images and high signal intensity on T_2-weighted images. They enhance brightly with gadolinium or with MRA.[12-14]

Solitary and Multiple Tumors. Glomus tumors have solitary and multiple forms. Solitary tumors are more common. Multiple glomus tumors make up only 10% of cases.[2] Solitary glomus tumors have been shown to occur at multiple locations within the same digit and in adjacent digits. Our case report demonstrates that solitary glomus tumors can occur simultaneously in nonadjacent digits. Compared with solitary glomus tumors, multiple tumors usually occur at an earlier age and are more common in males. Multiple glomus tumors are inherited in an autosomal-dominant pattern with incomplete penetrance and have been associated with neurofibromatosis type 1.[15] Clinically, multiple tumors are less likely to be painful and usually occur on the trunk and upper extremities. The malignant form of glomus tumor is glomangiosarcoma. Glomangiosarcomas are extremely rare and can arise within a typical glomus tumor or can arise de novo. De novo glomangiosarcomas are the only type that has been shown to metastasize.[16-18]

Histology. The histologic diagnosis of glomus tumor is based on the types of cellular tissue present and the degree of vascularity within the tumor. Type I, mucoid

hyaline, has hyalinized cartilage within islands of glomus cells. Type II, solid, consists of masses of glomus cells with very little vascularity; type II is commonly referred to as *glomus tumor* and is most common in solitary glomus tumors. Type III, angiomatous, has increased vascularity and is referred to as *glomangioma*; type III is usually present in patients with multiple glomus tumors, though these patients can have several tumors of different histologic types at the same time.[19]

Treatment and Outcome

The treatment for a symptomatic glomus tumor is marginal excision. When the tumor is excised with negative margins, this treatment is curative. For excision of digital glomus tumors, there are 2 surgical approaches—transungual and lateral subperiosteal.[2]

The more common, transungual approach involves removing the nail plate and making a longitudinal incision within the nail bed. Glomus tumors are generally easily released from the surrounding subungual tissue. One advantage of the transungual approach is that wide exposure of the nail bed allows for definitive visualization of the tumor margins and identification of additional glomus tumors within the nail bed. After tumor removal, the nail bed is repaired, and the nail plate may be replaced under the eponychial fold as a biologic dressing.

The lateral subperiosteal approach involves making an incision dorsal to the midlateral line of the finger. The dissection is made down to the distal phalanx, and a subperiosteal flap is raised dorsally. The glomus tumor is encountered within the flap and excised. The flap is then sutured back into its original position and closed. Fewer postoperative nail deformities exist with the lateral subperiosteal approach.[20]

Regardless of which approach is used, recurrence rates in the digits range from 5% to 50%.[1,20,21] Higher recurrence rates are related to incomplete excision of the original tumor or to another tumor going unrecognized at time of initial excision. If the recurrence occurs within weeks to months, it is the result of inadequate excision; if it is delayed for years, it is likely the result of a new solitary glomus tumor.[19]

Conclusions

Our patient's case shows that multiple solitary glomus tumors must be included in the differential diagnosis for a patient with pain in multiple fingers. The literature indicates that, when multiple solitary tumors occur, their sizes are usually different. Therefore, when a glomus tumor is found, it is important to examine the subungual and paraungual surfaces of all the fingers to determine if multiple solitary glomus tumors are present. This is particularly important in patients who are undergoing surgical excision of a glomus tumor. Larger case studies are needed to determine the exact incidence of multiple solitary glomus tumors.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

"...when a glomus tumor is found, it is important to examine the subungual and paraungual surfaces of all the fingers to determine if multiple solitary glomus tumors are present."

References

1. Carrol RE, Berman AT. Glomus tumors of the hand: review of the literature and report on twenty-eight cases. *J Bone Joint Surg Am.* 1972;54(4):691-703.
2. Rettig AC, Strickland JW. Glomus tumor of the digits. *J Hand Surg Am.* 1977;2(4):261-265.
3. Posch JL. Tumors of the hand. *J Bone Joint Surg Am.* 1956;38(3):517-540.
4. Shugart RR, Soule EH, Johnson EW. Glomus tumor. *Surg Gynecol Obstet.* 1963;117:334-340.
5. Greene RG. Soft tissue tumors of the hand and wrist: a 10 year survey. *J Med Soc NJ.* 1964;61:495-498.
6. Kale SS, Roa VK, Bentz ML. Glomus tumor of the index finger. *J Craniofac Surg.* 2006;17(4):801-804.
7. Giele H. Hildreth's test is a reliable clinical sign for the diagnosis of glomus tumours. *J Hand Surg Br.* 2002;27:157-158.
8. Ekin A, Ozkan M, Kabaklioglu T. Subungual glomus tumours: a different approach to diagnosis and treatment. *J Hand Surg Br.* 1997;22(2):228-229.
9. Bhaskaranand K, Navadgi BC. Glomus tumour of the hand. *J Hand Surg Br.* 2002;27(3):229-231.
10. Hou SM, Shih TT, Lin MC. Magnetic resonance imaging of an obscure glomus tumour in the finger tip. *J Hand Surg Br.* 1993;18(4):482-483.
11. Fornage BD. Glomus tumors in the fingers: diagnosis with US. *Radiology.* 1988;167(1):83-85.
12. Jablon M, Horowitz A, Bernstein DA. Magnetic resonance imaging of a glomus tumor of the fingertip. *J Hand Surg Am.* 1990;15(3):507-509.
13. 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(3):472-475.
14. Theumann NH, Goettmann S, Le Viet D, et al. Recurrent glomus tumors of fingertips: MR imaging evaluation. *Radiology*. 2002;223(1):143-151.
15. Moor EV, Goldberg I, Westreich M. Multiple glomus tumor: a case report and review of the literature. *Ann Plast Surg.* 1999;43(4):436-438.
16. Plate A, Lee SJ, Steiner G, Posner M. Tumorlike lesions and benign tumors of the hand and wrist. *J Am Acad Orthop Surg.* 2003;11(2):129-141.
17. Aiba M, Hirayama A, Kuramochi S. Glomangiosarcoma in a glomus tumor: an immunohistochemical and ultrastructural study. *Cancer.* 1988;61(7):1467-1471.
18. Hiruta N, Kameda N, Tokudome T, et al. Malignant glomus tumor: a case report and review of the literature. *Am J Surg Pathol.* 1997;21(9):1096-1103.
19. Tuncali D, Yilmaz AC, Terzioglu A, Aslan G. Multiple occurrences of different histologic types of glomus tumor. *J Hand Surg Am.* 2005;30(9):161-164.
20. Vasisht B, Watson HK, Joseph E, Lionelli GT. Digital glomus tumors: a 29-year experience with a lateral subperiosteal approach. *Plast Reconstr Surg.* 2004;114(6):1486-1489.
21. Van Geertruyden J, Lorea P, Goldschmidt D, et al. Glomus tumours of the hand. A retrospective study of 51 cases. *J Hand Surg Br.* 1996;21(2):257-260.

This paper will be judged for the Resident Writer's Award.