

Large Solitary Glomus Tumor of the Wrist Involving the Radial Artery

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Abstract

Glomus tumors are neuromyoarterial glomus body neoplasms that most commonly occur in the subungual areas of the hand. These tumors typically present as painful solitary lesions with localized tenderness. Extradigital glomus tumors are rare and difficult to diagnose because of their varying symptoms and presentation. Misdiagnosis can result in significant chronic pain, disuse syndromes, and disability over prolonged periods. Timely diagnosis and surgical resection are generally curative with complete resolution of symptoms. Surgeons should be aware of this rare condition and consider it in the differential diagnosis when treating a painful soft-tissue mass of the wrist.

We report a case of a large solitary glomus tumor that occurred on the volar-radial aspect of the wrist and involved the radial artery. The patient was successfully treated with complete surgical excision of the tumor. There were no symptoms or recurrence at 3-year follow-up. In the literature, this is the first case of large atypical wrist glomus tumor involving the radial artery to be reported with corresponding magnetic resonance imaging, intraoperative, histology, and immunohistochemistry findings.

Glomus tumors are neoplasms that originate from normal glomus bodies in the skin and are most commonly found in the subungual areas of the digits.¹ Glomus bodies are neuromyoarterial structures in the reticular dermis that serve as specialized arteriovenous anastomoses. These bodies contain afferent arterioles and efferent veins with multiple connections, and glomus cells have contractile properties because of their similarity to smooth muscle cells.^{1,2} Glomus bodies help regulate blood flow and temperature of the skin and are found in their largest concentration in the fingertips, palms of the hands, and soles of the feet.^{3,4}

Glomus tumors represent hyperplastic glomus bodies and make up 1% to 4.5% of upper extremity neoplasms, with

approximately 75% in the hand and 50% in the subungual area.^{1,5,6} These tumors can also present in multiple locations at once and can occur in atypical and ectopic locations.³ Although generally benign, glomus tumors can also exhibit malignant and metastatic potential in rare cases.^{7,8} They can also be locally aggressive with bony destruction of the distal phalynx.^{2,9,10} Tumors typically present as painful solitary soft-tissue lesions that are exquisitely tender to palpation, dark red-purple or bluish, and hypersensitive to cold.^{5,10} Van Geertruyden and colleagues¹⁰ reported that the diagnosis of glomus tumor can be made clinically in 90% of cases. However, glomus tumors can easily be mistaken for other lesions, such as hemangiomas, angiomas, neuromas, neurofibromas, lipomas, and ganglion cysts. An inaccurate or incomplete workup can result in persistent pain and symptoms along with intraoperative complications.³ Magnetic resonance imaging (MRI), the most sensitive imaging modality for detecting glomus tumors of the hand, can assist in the workup.^{3,11,12}

Extradigital glomus tumors are difficult to diagnose because of their rarity and unspecific symptoms and presentation.¹³ Misdiagnosis and delayed diagnosis can result in significant chronic pain, disuse syndromes, and disability.^{1,10} Correct diagnosis and surgical resection are generally curative with complete resolution of symptoms.

In this article, we report a case of a large atypical glomus tumor that occurred on the wrist and involved the radial artery. This tumor was successfully treated with surgical excision. The patient provided written informed consent for print and electronic publication of this case report.

Case Report

A 63-year-old man presented to clinic with an extremely tender soft-tissue mass on his nondominant, left wrist. The mass had been increasing in size for a year. It was painless at rest but very painful to light palpation, with referred pain proximally up to the shoulder.

The patient did not recall any traumatic or inciting event, had not undergone any prior workup or treatment for symptoms, and had no history of masses elsewhere on the body. Past medical history was significant for type 2 diabetes and colon and prostate cancer, which had been treated with che-

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Figure 1. Soft-tissue mass on volar-radial aspect of left wrist proximal to thenar eminence and radial to flexor carpi radialis tendon.

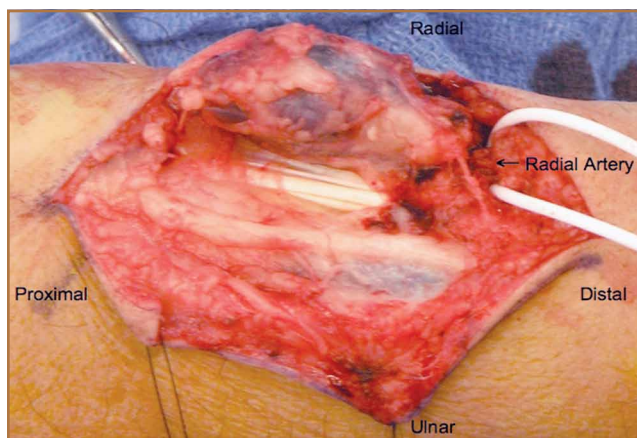


Figure 3. Intraoperative photograph of bluish mass on volar-radial aspect of left wrist shows relationship to radial artery. Distal aspect of radial artery is retracted with white Vesselloop (Argon Medical, Plano, Texas).

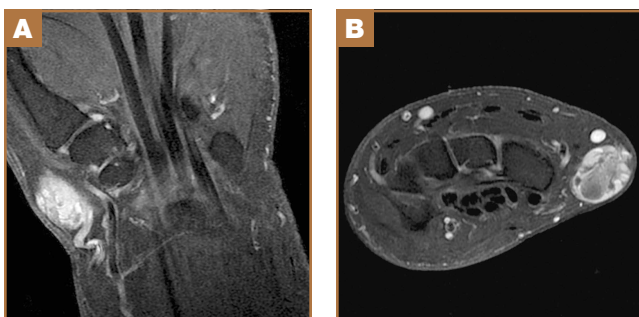


Figure 2. T2-weighted (A) coronal and (B) axial magnetic resonance imaging shows heterogeneous subcutaneous mass adjacent to radial artery with increased signal intensity from vascular feeding vessels.

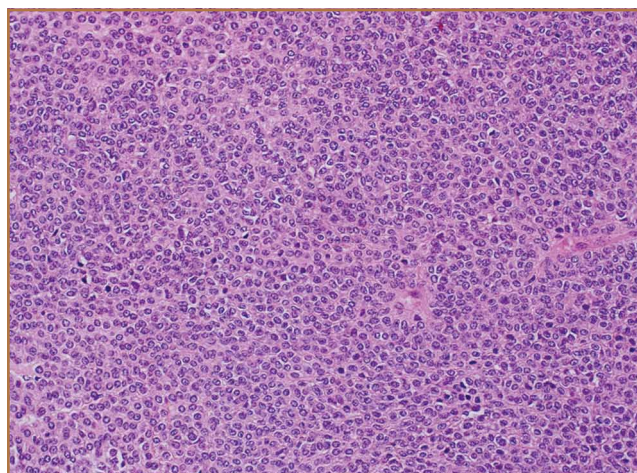


Figure 4. Histology of mass shows solid sheets of uniform round cells with interspersed capillaries and centrally placed nuclei (hematoxylin-eosin, original magnification $\times 250$).

motherapy and was now in remission.

Physical examination revealed a 2×2.5-cm well-circumscribed soft-tissue mass on the volar-radial aspect of the left wrist proximal to the thenar eminence and radial to the flexor carpi radialis tendon (**Figure 1**). The mass was soft, mobile, and nonfluctuant and did not transilluminate. The overlying skin was normal in color and appearance—no discoloration, erythema, wounds, or drainage. The radial artery was palpable, and the mass did not pulsate or have a bruit. The patient had normal wrist range of motion limited by pain on compression of the mass with motor and sensation intact throughout the hand. Plain radiographs of the wrist showed no bony pathology or involvement from the mass. A soft-tissue shadow was visible around the wrist without calcifications. A wrist MRI was performed to better evaluate the mass, and the T2-weighted images showed a heterogeneous subcutaneous mass adjacent to the radial artery with increased signal intensity from surrounding feeding vessels (**Figure 2**).

Given the clinical and imaging findings, there was concern for a possible vascular tumor. Therefore, excisional biopsy was recommended over needle biopsy because of the bleeding risk. With the patient under general anesthesia, and a tourniquet

used without exsanguination, a Brunner-type zigzag incision was made centered over the mass with elevated skin flaps. The 2.7×2.6×1.1-cm mass was superficial and involved the radial artery (**Figure 3**). After the radial artery was dissected proximally and distally, 2 perforating vessels were found entering the mass. These vessels were ligated, which allowed the mass to be peeled completely off the artery. Histology with hematoxylin-eosin staining showed solid sheets of uniform round cells with interspersed capillaries and centrally placed nuclei without evidence of malignancy (**Figure 4**).

The tourniquet was released before skin closure, and adequate hemostasis was obtained. The wound was closed, and the patient was placed in a volar wrist splint for immobilization. Pain relief after excision of the mass was immediate, and the postoperative course uneventful. After surgery, immunohistochemistry of the mass showed minimal mitotic activity,

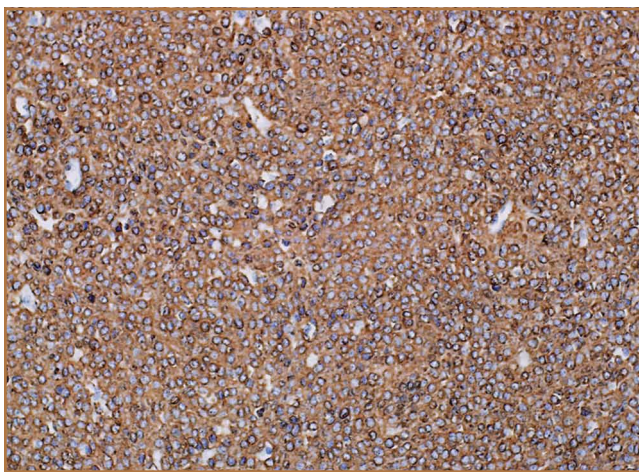


Figure 5. Immunohistochemistry shows positive staining for smooth muscle actin, confirming diagnosis of glomus tumor (immunoperoxidase, original magnification $\times 250$).

with a positive immunoperoxidase stain for smooth muscle actin confirming a diagnosis of glomus tumor (**Figure 5**). At 3-year follow-up, the patient had no pain, symptoms, or tumor recurrence.

Discussion

Glomus tumors are an established cause of pain in the subungual areas of the hand; numerous cases have been reported.^{1,5,10,14} However, extradigital glomus tumors, particularly those involving the wrist, are rare, and only a few have been described. Given the lack of consistent findings and presentations, diagnosis is difficult. Case series have documented an overall 2:1 female-to-male predominance of glomus tumors,⁶ but extradigital tumors are more common in men (4.6:1 male-to-female ratio).³ Extradigital glomus tumors are commonly diagnosed between ages 40 and 80 years. Classic symptoms of subungual tumors include pain, localized tenderness, and cold hypersensitivity,^{1,10} but symptoms are much more variable with extradigital locations. Previous trauma or injury to the lesion area is reported in 20% to 30% of cases before symptom onset.^{3,15} Intravascular locations of glomus tumors are extremely rare; only 4 cases of tumors involving venous structures have been reported.¹⁶⁻¹⁹ In the present case, the patient's main complaints were pain and localized tenderness associated with a progressively increasing mass without any history of trauma. The large size of his mass (~2.5 cm in diameter) on examination was unique, as was involvement of the radial artery.

Misdiagnosis and delayed diagnosis of extradigital glomus tumors are common, and symptoms such as chronic pain typically persist for 7 to 11 years before the correct diagnosis is made.^{1,10} On average, 2.5 physician consultants (including psychiatrists) evaluate the patient before glomus tumor is identified.¹⁰ There are other reports of atypical or ectopic glomus tumors taking 5 to 25 years to be diagnosed.²⁰⁻²² The differential diagnosis for glomus tumors includes hemangiomas, cellular or cavernous hemangiomas, vascular tumors, neuro-

mas, neurofibromas, lipomas, paragangliomas, ganglion cysts, pigmented nevi, Pacinian corpuscle hyperplasia, and foreign bodies. A key element of clinical diagnosis is the disproportionate amount of pain and localized tenderness caused by the lesion relative to its size. The hypersensitivity of this tumor is thought to result from enlargement of the tumor and impingement on nearby Pacinian corpuscles, nerve endings in the skin that are responsible for sensitivity to vibration and pressure.^{2,9}

Plain radiographs can be useful in detecting glomus tumors of the hand but are less helpful with extradigital tumors, with identification rates of 24% in certain series.³ MRI is the most sensitive imaging modality for diagnosing glomus tumors of the hand; a detection rate of 80% to 100% has been reported in various case series.^{3,11,12} Specificity of MRI for glomus tumors has been reported at 50%.^{11,23} Placement of a radiographic marker directly over the area of most pain can assist in tumor localization.³ Glomus tumors typically have decreased signal intensity on T1-weighted images and increased intensity on T2-weighted images, but signal patterns are variable and particularly difficult to differentiate with small tumors. MRI is useful in the setting of recurrent glomus tumors, where incomplete excision is possible. In 24 cases of continued pain after glomus tumor excision, Theumann and colleagues²⁴ used MRI to identify a nodule consistent with recurrent glomus tumor in all patients. Three-dimensional contrast-enhanced magnetic resonance angiography (MRA) can also help diagnose glomus tumors while providing valuable information regarding size and location for surgical planning.^{25,26} With MRA, it is crucial to evaluate the arterial or arteriovenous phase of imaging, as the glomus tumor is richly vascularized and shows contrast enhancement after intravenous injection of gadolinium.²⁷ Angiography, ultrasonography, thermography, and scintigraphy have all been used to diagnose glomus tumors but have shown limited utility and accuracy.¹¹

Treatment of glomus tumors is complete surgical excision because of their relatively small size and subcutaneous location. Resection success rates are consistently higher than 95%, with resolution of all symptoms.^{1,10,14} Local recurrence of tumors after excision occurs in 1% to 33% of cases, depending on series, and may be immediate or delayed, with immediate recurrence commonly caused by inadequate excision.^{1,10,15,28} Delayed recurrence is less common and presents several years after excision, typically with a new growth near the previous excision.¹⁰ Recurrence years after surgery may also represent multiple tumors unrecognized during initial workup and can be treated with repeat excision or radiotherapy.

Robert and colleagues²⁹ recently reported the case of a glomus tumor, on the dorsal aspect of the wrist, discovered incidentally in a 71-year-old patient and treated with surgical excision. Several years earlier, Chim and colleagues³⁰ described a similar case, of a large wrist glomus tumor worked up with MRI. In a retrospective review of all extradigital glomus tumors seen over a 20-year period, Schiefer and colleagues³ reported 4 glomus tumors of the wrist out of 56 tumors total. The most common sites were forearm (11 cases) and knee (10 cases), and the majority of patients presented with pain and localized

tenderness. Mean tumor size was 0.66 cm (range, 0.1-0.3 cm), with 77% of tumors less than 1 cm. Our patient's 2.7×2.6×1.1-cm tumor was large for a glomus tumor. Its involvement with the radial artery feeding vessels likely contributed to its large and progressively increasing size. It is worth noting that, in the series by Schiefer and colleagues,³ the only patient with symptoms persisting after excision had a large (3 cm in diameter) deep tumor of the foot; the entire tumor was removed, and there was no recurrence by 10-year follow-up. Folpe and colleagues⁷ suggested that deep tumors larger than 2 cm should be at higher suspicion for malignancy. Joseph and Posner²¹ reported 3 cases of glomus tumors, on the ulnar side of the wrist, diagnosed with help of a provocative test using ethyl chloride spray.

Conclusion

Overall, glomus tumors are rare and challenging to diagnosis and should be in the differential in any symptomatic patient with a painful soft-tissue mass of the wrist. Advanced imaging studies, such as MRI, can assist in localization, diagnosis, and preoperative planning. Histology and immunohistochemistry are essential to differentiate glomus tumor from other vascular tumors, and complete excision is necessary to prevent local recurrence.

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