

# Blue Nodules on the Forearms in an Active-Duty Military Servicemember

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A 31-year-old active-duty military servicemember presented to the dermatology clinic for evaluation of 0.3- to 2-cm, tender, blue nodules on the wrists and forearms. The lesions first appeared on the right wrist secondary to a presumed injury sustained approximately 10 years prior to presentation and spread to the proximal forearm as well as the left wrist and forearm. He denied fevers, chills, chest pain, hematochezia, hematuria, or other skin findings. Physical examination revealed blue-violaceous, firm nodules on the right volar wrist and forearm that were tender to palpation. Blue-violaceous, papulonodular lesions on the left volar wrist and dorsal hand were not tender to palpation. A punch biopsy was performed.

## WHAT'S YOUR DIAGNOSIS?

- cutaneous leiomyoma
- glomangiomyoma
- hereditary hemorrhagic telangiectasia
- Kaposi sarcoma
- neurilemmoma

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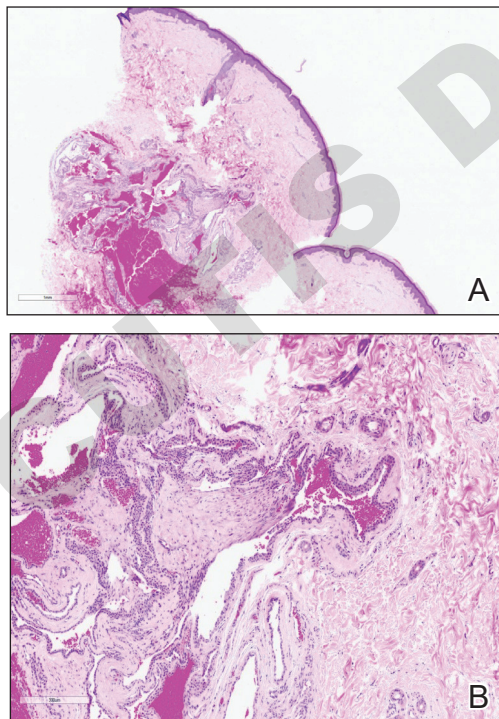
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## THE DIAGNOSIS: Glomangiomyoma

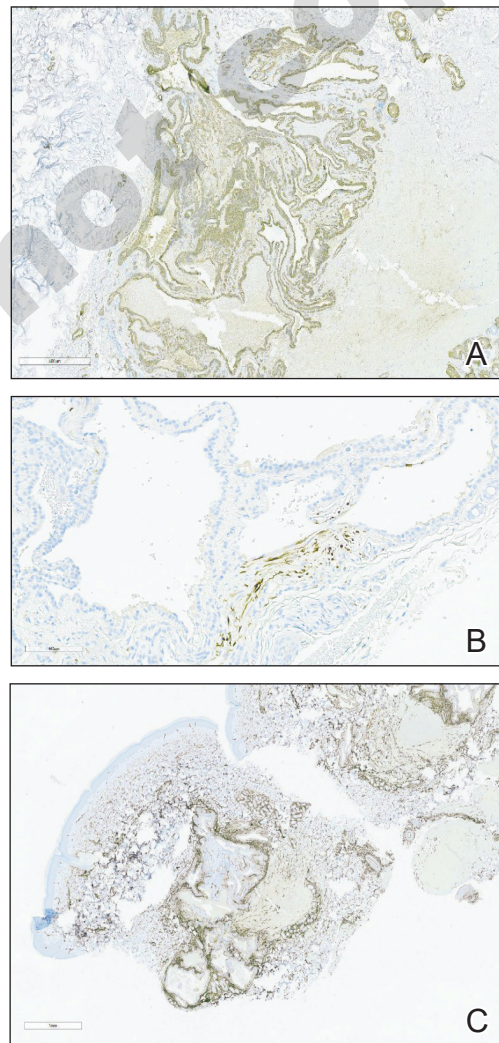
A punch biopsy of the right forearm revealed a collection of vascular and smooth muscle components with small and spindled bland cells containing minimal eosinophilic cytoplasm (Figure 1), confirming the diagnosis of glomangiomyoma. Immunohistochemical stains also supported the diagnosis and were positive for smooth muscle actin, desmin, and CD34 (Figure 2). Magnetic resonance imaging from a prior attempt at treatment with sclerotherapy demonstrated scattered vascular malformations with no notable internal derangement. There was no improvement with sclerotherapy. Given the number and vascular nature of the lesions, a trial of pulsed dye laser (PDL) therapy was administered and tolerated by the patient. He subsequently moved to a new military duty station. On follow-up, he reported no noticeable clinical improvement in the lesions after PDL and opted not to continue with laser treatment.

Glomangiomyoma is a rare and benign glomus tumor variant that demonstrates differentiation into the smooth muscle and potentially can result in substantial complications.<sup>1</sup> Glomus tumors generally are benign neoplasms

of the glomus apparatus, and glomus cells function as thermoregulators in the reticular dermis.<sup>2</sup> Glomus tumors comprise less than 2% of soft tissue neoplasms and generally are solitary nodules; only 10% of glomus tumors occur with multiple lesions, and among them, glomangiomyoma is the rarest subtype, presenting in only 15% of cases.<sup>2,3</sup> The 3 main subtypes of glomus tumors are solid, glomangioma, and glomangiomyoma.<sup>4</sup> Clinically, the lesions may present as small blue nodules with associated pain and cold or pressure sensitivity.



**FIGURE 1.** A and B, Histopathology revealed a collection of dilated and variably sized vascular spaces in the dermis surrounded by small bland cells with little cytoplasm as well as some foci between the vascular spaces containing cells that were more spindled and had increased amounts of eosinophilic cytoplasm (H&E, original magnifications  $\times 20$  and  $\times 80$ ). Reference bars indicate 1 mm and 300  $\mu\text{m}$ , respectively.



**FIGURE 2.** A, Immunohistochemistry revealed small cells that were decorated with smooth muscle actin (original magnification  $\times 50$ ). B, Spindled cells were highlighted with desmin (original magnification  $\times 200$ ). C, CD34 highlighted the endothelial cells lining the spaces (original magnification  $\times 20$ ). Reference bars indicate 100  $\mu\text{m}$ , and 1 mm, respectively.

Although there appears to be variation of the nomenclature depending on the source in the literature, glomangiomas are characterized by their predominant vascular malformations on biopsy. Glomangiomyomas are a subset of glomus tumors with distinct smooth muscle differentiation.<sup>4</sup> Given their pathologic presentation, our patient's lesions were most consistent with the diagnosis of glomangiomyoma.

The small size of the lesions may result in difficulty establishing a clinical diagnosis, particularly if there is no hand involvement, where lesions most commonly occur.<sup>2</sup> Therefore, histopathologic evaluation is essential and is the best initial step in evaluating glomangiomyomas.<sup>4</sup> Biopsy is the most reliable means of confirming a diagnosis<sup>2,4,5</sup>; however, diagnostic imaging such as a computed tomography also should be performed if considering blue rubber bleb nevus syndrome due to the primary site of involvement. Surgical excision is the treatment of choice after confirming the diagnosis in most cases of symptomatic glomangiomyomas, particularly with painful lesions.<sup>6</sup>

Neurilemmomas (also known as schwannomas) are benign lesions that generally present as asymptomatic, soft, smooth nodules most often on the neck; however, they also may present on the flexor extremities or in internal organs. Although primarily asymptomatic, the tumors may be associated with pain and paresthesia as they enlarge and affect surrounding structures. Neurilemmomas may occur spontaneously or as part of a syndrome, such as neurofibromatosis type 2 or Carney complex.<sup>7</sup>

Hereditary hemorrhagic telangiectasia (formerly known as Osler-Weber-Rendu syndrome) is an autosomal-dominant disease that presents with arteriovenous malformations and telangiectases. Patients generally present in the third decade of life, with the main concern generally being epistaxis.<sup>8</sup>

Kaposi sarcoma is a viral infection secondary to human herpesvirus 8 that results in red-purple lesions commonly on mucocutaneous sites. Kaposi sarcoma can be AIDS associated and non-HIV associated. Although clinically indistinguishable, a few subtle histologic features can assist in differentiating the 2 etiologies. In addition to a potential history of immunodeficiency, evaluating for involvement of the lymphatic system, respiratory tract, or gastrointestinal tract can aid in differentiating this entity from glomus tumors.<sup>9</sup>

Leiomyomas are smooth muscle lesions divided into 3 subcategories: angioleiomyoma, piloleiomyoma, and genital leiomyoma. The clinical presentation and histopathology will vary depending on the subcategory. Although cutaneous leiomyomas are benign, further workup for piloleiomyoma may be required given the reported association with hereditary leiomyomatosis and renal cell cancer (Reed syndrome).<sup>10</sup>

Imaging can be helpful when the clinical diagnosis of a glomus tumor vs other painful neoplasms of the skin is unclear, such as in blue rubber bleb nevus syndrome, angioleiomyomas, neuromas, glomus tumors, leiomyomas, eccrine spiradenomas, congenital vascular malformations, schwannomas, or hemangiomas.<sup>4</sup> Radiologic findings for

glomus tumors may demonstrate cortical or cystic osseous defects. Magnetic resonance imaging and ultrasonography can help provide additional information on the lesion size and depth of involvement.<sup>1</sup> Additionally, deeper glomangiomyomas have been associated with malignancy,<sup>2</sup> potentially highlighting the benefit of early incorporation of imaging in the workup for this condition. Malignant transformation is rare and has been reported in less than 1% of cases.<sup>6</sup>

Treatment of glomus tumors predominantly is directed to the patient's symptoms; asymptomatic lesions may be monitored.<sup>4</sup> For symptomatic lesions, therapeutic options include wide local excision; sclerotherapy; and incorporation of various lasers, including Nd:YAG, CO<sub>2</sub>, and flash-lamp tunable dye laser.<sup>4,5</sup> One case report documented use of a PDL that successfully eliminated the pain associated with glomangiomyoma; however, the lesion in that report was not biopsy proven.<sup>11</sup>

Our case highlights the need to consider glomus tumors in patients presenting with multiple small nodules given the potential for misdiagnosis, impact on quality of life with associated psychological distress, and potential utility of incorporating PDL in treatment. Although our patient did not report clinical improvement in the appearance of the lesions with PDL therapy, additional treatment sessions may have helped,<sup>11</sup> but he opted to discontinue. Follow-up for persistently symptomatic or changing lesions is necessary, given the minimal risk for malignant transformation.<sup>6</sup>

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