

# A case of a glomus tumor presenting as an atypical hyperkeratotic papule of the hypothenar palm



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**Key words:** benign neoplasm; hypothenar eminence; glomus tumor; vascular neoplasm.

## INTRODUCTION

Glomus tumors are rare, benign vascular neoplasms typically localized to the digits, with a propensity for the subungual region of the hand. These subcutaneous hamartomas are derived from hyperplastic modified smooth muscle cells, specifically from the thermoregulatory glomus body found at the reticular dermis.<sup>1</sup> Clinical characteristics include a triad of severe focal paroxysmal pain, pinpoint tenderness, and cold sensitivity, often presenting as a raised red or blue-colored, painful nodule at acral or subungual areas.<sup>2-4</sup> However, glomus tumors are also reported in extradigital locations, especially in men.<sup>5,6</sup> The low incidence and varying presentations of glomus tumors pose a diagnostic challenge, leading to delayed diagnosis and unnecessary pain. Here we describe a patient with an atypical clinical presentation of a glomus tumor, uniquely located at the hypothenar eminence of the hand and resembling a foreign body granuloma.

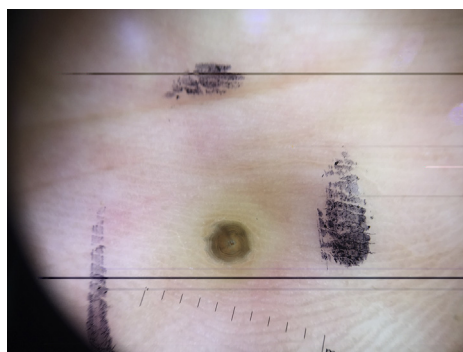
## CASE REPORT

A healthy 55-year-old Bahamian woman presented with a raised, painful hyperkeratotic brown papule that was on her right hand for 1 year. The pain was intermittent and sharp, aggravated with hand movement, and relieved with rest. At the time of the presentation, the patient lived in the Bahamas and did not indicate whether warm or cold temperatures made the pain worse. The patient denied any prior trauma.

On examination, she had a single 2-mm tender, hyperkeratotic brown papule at the hypothenar eminence of her right hand (Fig 1). Dermoscopy



**Fig 1.** A glomus tumor of the hypothenar eminence. Photo from clinical examination shows a hyperkeratotic papule. Note the resemblance of a foreign body.



**Fig 2.** Glomus tumor viewed with dermoscopy shows brown concentric rings.

found a lesion with concentric rings (Fig 2). Skin biopsy findings showed a circumscribed intradermal proliferation of round cells with vesicular nuclei without mitotic activity. Focal admixed vessels were

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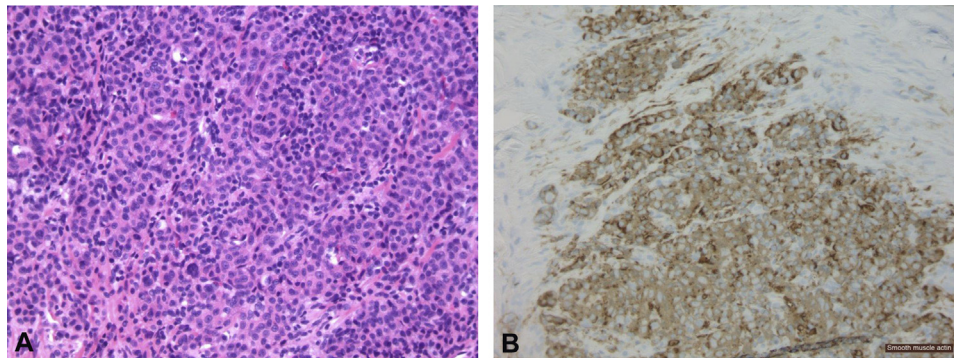
Conflicts of interest: None declared.

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**Fig 3.** **A**, Round cells proliferating intradermally, with vesicular nuclei and without mitotic activity. Admixed vessels were identified (H&E stain). **B**, Immunohistochemical stain for smooth muscle actin.

identified. Immunohistochemical staining was positive for smooth muscle actin and negative for S100 protein, suggestive of a glomus tumor (Fig 3). The patient was recommended to have wide local excision. When contacted, she reported continued intermittent stabbing daily pain in the area. Unfortunately, the patient did not follow up with the recommended treatment.

## DISCUSSION

Glomus tumors are rare, subcutaneous benign neoplasms typically found at the subungual area or at the pulp of the finger. Neuromyoarterial glomus bodies are contractile structures involved in thermoregulation that are found at the fingers, palms, and soles but are most highly concentrated at the nail bed.<sup>5</sup> Some literature, however, indicates that extradigital glomus tumors are more common than those found at the digits. A 20-year review of cases at the Mayo Clinic found 61% of all glomus tumors were extradigital.<sup>7</sup> The unsuspecting high frequency of extradigital locations contributes to missed diagnoses. Despite effective treatment options, patients have an average of 7 years of unrelieved pain before the condition is correctly diagnosed.<sup>2,5</sup> Glomus tumors have not been reported at the hypothenar eminence in the literature, although a few cases have been localized to the hands and wrists.<sup>1,2,8</sup>

The histologic features of glomus tumors include a variable composition of glomus cells, blood vessels, and smooth muscle cells. Based on these features, glomus tumors are classified into 3 types: solitary, glomangioma, or glomangiomyoma. The most common form is a solitary glomus tumor at the nail bed, composed of both vasculature and smooth muscle cells.<sup>7</sup> Other variants include a predominantly vascular glomangioma or a glomangiomyoma, which contains proportionate amounts of vascular and smooth muscle cells.<sup>3</sup> Interestingly,

extradigital glomus tumors are more likely to be glomangiomas.<sup>6</sup> Fortunately, malignant glomus tumors are rare.<sup>1,3,6</sup> Highly cellular glomus tumors have been mistaken for intradermal nevi, but S100 stain helps to distinguish whether the lesion is of melanocytic origin.<sup>3</sup> S100 was negative in our patient, excluding the differential of melanocytic nevus.

The cause of glomus tumors is unknown, but subungual tumors have been associated with neurofibromatosis and reactive hyperplasia secondary to trauma.<sup>1,8</sup> Multiple glomus tumors are painless and less common than solitary glomus tumors, but are hereditarily linked to chromosome 1p21-22 and mutated glomulin gene that has an unknown function.<sup>1,3</sup> Several hypotheses explain the pathogenesis of pain from a glomus tumor. Vasodilation of the glomus body triggered by cold temperatures may cause pain, which may be exacerbated by pressure. Mast cell degranulation that hypersensitizes thermal receptors may also contribute to pain. Nonmyelinated nerve fibers within glomus tumors are another potential source of pain.<sup>1</sup>

The key diagnostic factor of a glomus tumor is a group of symptoms, referred to as the *Carroll triad*, which includes paroxysmal pain, an extreme stabbing sensation, and cold hypersensitivity.<sup>1,3,8</sup> Numerous diagnostic methods have been used including plain radiography, Doppler ultrasonography, magnetic resonance imaging, and highly sensitive physical examination maneuvers such as the Love pin test, the Hildreth test, or the cold hypersensitivity test.<sup>8</sup> The Love pin test identifies the exact location of a glomus tumor by applying pinpoint pressure to the tender area. The Hildreth test removes pain by applying a tourniquet to the affected upper extremity to induce ischemia. The cold sensitivity test elicits increased pain while submerging the affected area in icy water.<sup>4,8</sup> In our case,

these tests were not performed because initially the suspicion for a glomus tumor was low based on its clinical appearance.

Overall, these benign growths are uncommon, often requiring a high index of suspicion, supported by clinical examination and skin biopsy, to be properly diagnosed. Glomus tumors in atypical locations can pose a particular challenge. Thus, being aware of the varying clinical presentations is important to streamline the diagnosis. Focusing on the classic symptoms of localized pain and cold sensitivity may aid in identifying glomus tumors. By promoting awareness of the typical and less common presentations of this unusual tumor, providers can properly identify and treat glomus tumors in a timely manner, thus improving patient quality of life.

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