

# Glomangioma: A Case Report and Review of the Literature

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## GOAL

To understand glomangiomas to better manage patients with these tumors

## LEARNING OBJECTIVES

Upon completion of this activity, dermatologists and general practitioners should be able to:

1. Describe the clinical and histologic appearance of glomus tumors, specifically glomangiomas.
2. Distinguish between lesions of blue rubber bleb nevus syndrome and glomangiomas.
3. Evaluate treatment options for glomangiomas.

## INTENDED AUDIENCE

This CME activity is designed for dermatologists and generalists.

CME Test on page 28.

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*Glomus tumors are benign localized tumors of the skin accounting for 1% to 2% of all soft tissue*

*tumors. They may present as a solitary tumor or multiple tumors, termed glomangioma. We describe a 69-year-old man with a medical history of psoriasis and hypertension who presented with an incidental finding of multiple asymptomatic, noncompressible, blue lesions over his arms, chest, and back. The lesions, present since childhood, had never been subject to a workup. The patient had no history of gastrointestinal bleeding and no known family history of similar*

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lesions. Physical examination revealed multiple nontender, blue, subcutaneous nodules that were 1 to 2 cm in diameter and located on the bilateral arms, chest, and back. The diagnosis of glomangioma was made and no further treatment was indicated.

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### Case Report

A 69-year-old man with a medical history of psoriasis and hypertension presented with an incidental finding of multiple asymptomatic, noncompressible, blue

lesions over his arms, chest, and back. The lesions were present since childhood and had never been subject to a workup. He had no history of gastrointestinal bleeding and no known family history of similar lesions. His medications included amlodipine besylate, efalizumab, and valsartan.

Physical examination revealed multiple nontender, blue, subcutaneous nodules that were 1 to 2 cm in diameter and located on the bilateral arms, chest, and back (Figure 1). A punch biopsy was performed from the chest lesions. Histopathologic examination revealed dilated vascular



**Figure 1.** Multiple nontender, blue, subcutaneous nodules on the right forearm measuring 1 to 2 cm in diameter (A) and on the left upper back and shoulder (B).

channels (Figure 2) that were positive for endothelial factor VIII and CD34 and surrounded by glomus cells positive for smooth muscle actin (Figure 3). The diagnosis of glomangioma was made and no further treatment was indicated.

### Comment

Glomangiomas are benign localized tumors of the skin characterized by abnormal, smooth muscle–like glomus cells.<sup>1</sup> In 1924, Masson<sup>2</sup> described the neurovascular glomus, which he later renamed the *neurovascular glomus*, and its tumors. The term *glomangioma* was coined by Bailey<sup>3</sup> in 1935 and is currently applied to lesions with a wide vascular lumen, which are most commonly found in patients with multiple tumors.<sup>4</sup>

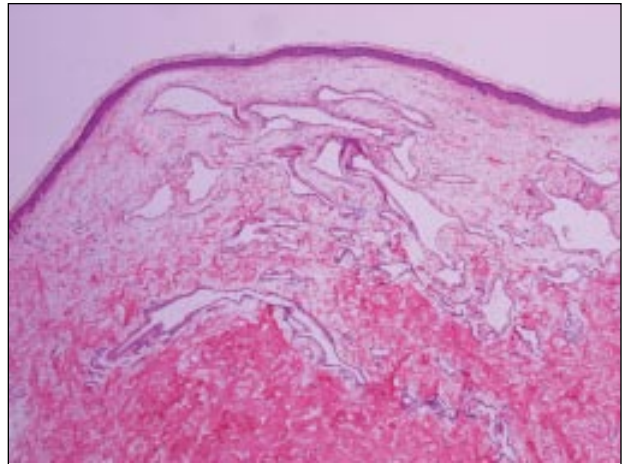
Glomus tumors arise from modified smooth muscle cells normally found in specialized arteriovenous shunts present in acral sites, especially the fingertips. This location reflects their function, as the arteriovenous anastomoses of these areas, also known as the Sucquet-Hoyer canals, are involved in temperature regulation. Sucquet-Hoyer canals are lined with endothelial cells, contain several layers of glomus cells in their walls, and connect afferent arterioles to efferent venules.<sup>5</sup>

Glomus tumors are thought to be hamartomas<sup>6</sup> and account for 1% to 2% of all soft tissue tumors.<sup>5</sup> There are 2 forms of glomus tumors, with the more common solitary variant accounting for 90% of cases and a more rare multiple variant, termed *glomangioma*, accounting for 10% of cases.<sup>7</sup>

The tumors of the solitary variant are small, painful, purple nodules with predilection for acral areas of the extremities, especially the nail beds of the fingers and toes.<sup>8</sup> Aching pain, well-localized tenderness, and temperature sensitivity are the characteristic triad of signs and symptoms.<sup>5</sup> They typically are less than 1 cm in diameter.<sup>8</sup>

In contrast, multiple glomus tumors are characterized as glomangiomas because of the angiomatous appearance of the lesions. Glomangiomas often appear during childhood as small bluish nodules situated deep in the dermis, widely scattered over the skin surface. They are rarely subungual and are less likely to be painful.<sup>8</sup> Glomangiomas have a predilection for the upper extremities and occasionally are found on the lower extremities, head, and back. They may be slightly larger and less well-circumscribed than solitary glomus tumors.<sup>5</sup>

An autosomal dominant inheritance pattern has been described for glomus tumors, with some types being mapped to band 11q23.<sup>6</sup> Familial cases have been reported with incomplete penetrance and variable expression.<sup>7</sup> Glomangiomas have a male



**Figure 2.** Histopathologic examination of a punch biopsy specimen reveals dilated vascular channels that are not specific to glomangioma and also can be found in blue rubber bleb nevus syndrome (H&E, original magnification  $\times 4$ ).

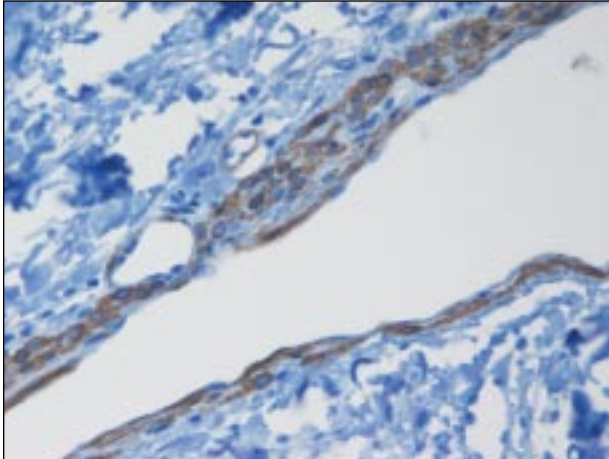
predominance, while females more frequently (in 70% of cases) are found to have solitary glomus tumors.<sup>5</sup>

Histopathologically, glomus tumors contain dilated vascular channels surrounded by glomus cells. The glomus cells are monomorphic round or polygonal cells with plump nuclei and scant eosinophilic cytoplasm. They are positive for smooth muscle actin, while vascular endothelium is positive for factor VIII and CD34.<sup>9</sup>

Choosing the appropriate treatment regimen for glomus tumors and glomangiomas should be individualized to the patient and guided by the clinical presentation. Treatment is not always indicated, particularly in asymptomatic cases of glomangioma. Surgical intervention, when needed, typically is excision with primary closure. Laser treatment, electromagnetic radiation, and sclerotherapy also have been used.<sup>5</sup>

*Blue Rubber Bleb Nevus Syndrome*—It is important to distinguish glomangioma from blue rubber bleb nevus syndrome (BRBNS), which is associated with venous malformations on both the skin and gastrointestinal tract. The BRBNS venous malformations of the gastrointestinal tract can be associated with clinically significant gastrointestinal bleeding.<sup>10</sup>

Lesions of BRBNS can be macular, papular, or nodular, and usually are multiple, varying in diameter from a few millimeters to several centimeters. The cutaneous lesions usually are asymptomatic and the overlying skin may show increased sweating. These lesions may appear at birth or in early childhood, and they tend to increase in size and frequency with age. Although they may occur anywhere, they are principally located on the upper limbs, trunk, and perineum. Acral lesions are unusual and the lesions have no evidence of malignant change.<sup>11</sup>



**Figure 3.** Punch biopsy results illustrate glomus cells, seen as navy blue cells within the dermal collagen. The glomus cells stain positive for smooth muscle actin, as does the lining of the dilated vascular channels. Glomus cells are specific to glomangioma, distinguishing it from blue rubber bleb nevus syndrome (smooth muscle actin, original magnification  $\times 400$ ).

Cutaneous lesions of BRBNS are blue, soft, and nipplelike, easily compressing and refilling slowly.<sup>12</sup> On the other hand, glomangiomas are noted for a distinct raised, often hyperkeratotic, cobblestone-like appearance, and could not be completely emptied by compression.<sup>13</sup> Glomangiomas generally do not extend into deep structures.<sup>14</sup>

Histologically, glomangiomas contain clusters of dilated vascular channels lined by a thin layer of endothelial cells in the dermis or subcutaneous fat. The walls are a fibrous stroma, occasionally containing smooth muscle.<sup>12</sup> Dilated vascular channels lined by endothelial cells are characteristic of both diseases. Therefore, biopsy results confirming the presence of glomus cells lining the dilated vascular channels characterize glomangiomas.<sup>10</sup>

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