A Rare Case of Extradigital Glomus Tumor

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Abstract

Glomus tumors are rare vascular tumors comprised of glomus cells surrounding capillary-sized vessels. These tumors are caused by a proliferation of cells from the glomus body, an anatomic unit primarily found in the hands, feet, and ears for thermoregulation. While the majority of tumors arise on the hands and under the fingernails, they can occur anywhere on the body. The tumors are exquisitely painful, but surgical excision is curative and provides instant pain relief. Here, we present a rare case of a solitary glomus tumor on the upper arm of a 77-year-old male patient.

Introduction

Glomus tumors are relatively rare lesions typically found underneath the fingernails of young adults. They are classically described as macules with extreme tenderness when palpated, purple/blue color, and distinct histopathology. When glomus cell lesions present as previously described, diagnosis is relatively straightforward. Glomus cell tumors on the fingers, however, tend to appear more exophytic or nodular. Lesions of this nature may mimic other dangerous cutaneous cancers and/or cancer metastases. We present a case of a glomus cell tumor on the upper arm of a patient with a history of infrequent medical care and several risk factors for visceral malignancy.

Case Presentation

A 77-year-old male presented to the clinic complaining of a painful nodule on his left upper arm. He said the lesion had been present for 10 years to 15 years but had begun to grow rapidly in the last three weeks. He complained that it had become substantially more painful recently.

Upon examination, the lesion was exquisitely tender to palpation, dark blue in color, exophytic, and nodular. It measured roughly 2.0 cm by 2.0 cm in diameter (Figure 1). The patient stated that he had not been to a doctor in more than 40 years nor completed any age-appropriate cancer screenings.

The upper-arm lesion was excised with narrow, 2-mm margins and sent for pathology. The differential diagnosis included: glomus cell tumor, Merkel cell carcinoma, leiomyoma, cancer metatasis, lymphoma/leukemia cutis, dermatofibrosarcoma protuberans, and undifferentiated pleomorphic sarcoma. Pathology later revealed a well-circumscribed, encapsulated dermal tumor (Figure 2, 2.5x) and blood vessels surrounded by sheets of round, monotonous glomus cells (Figure 3, 10x), confirming a rare case of extra-digital glomus cell tumor (Figure 3). The patient did not return for suture removal or follow-up after the excision.

Discussion

A glomus tumor is a benign proliferation of cells that originate from perivascular cells of the glomus body. The tumors tend to be small (< 1 cm to 2 cm), solitary papules with a violaceous tint and demonstrate extreme sensitivity to pain. The glomus body itself is an anatomic and functional unit utilized by the body for thermoregulation. The unit is comprised of an afferent arteriole leading into a Sucquet-Hoyer canal and an efferent venule with surrounding smooth muscle cells, glomus cells, and nerve fibers. The glomus body acts as a shunt to link the arteriole to the venule, therefore bypassing capillaries. These regulatory units are located all over the body but are found in significantly higher numbers in the pads of fingers and toes, nail beds, palms, soles, and thin skin of the ear. Hoyer first described the glomus unit in 1877, but the glomus neoplasm was not fully described until 1924 by Masson. What leads to glomus cell proliferation and tumor formation is unknown, although in one study, 20% to 30% of patients reported the tumors arose in areas of previous trauma.

Clinically, there are three main subtypes of tumors that contain glomus bodies: glomus tumors, glomangiomas, and glomangiomyomas. Solitary glomus tumors are the most common and represent 75% to 90% of all glomus neoplasms. They tend to arise during young adulthood (ages 20 to 40) and show no gender preference, with the exception that subungual lesions are more prominent in women. They typically present as 1-cm to 2-cm nodules with a violaceous tint that are tender to touch and extremely sensitive to temperature extremes and pressure. Histologically, the glomus tumor shows an abundant proliferation of round glomus cells with pale eosinophilic cytoplasm surrounding capillary-sized vascular structures. In contrast, glomangiomas tend to arise in childhood and consist of multiple small, red-blue tumors that are generally painless. Under the microscope, these tumors show venous malformation with few glomus cells. Glomangiomymomas demonstrate a predominance of smooth muscle cell proliferation. All three subtypes are considered benign.

The most common location of solitary glomus tumors is on the digits of the hand, typically subungual. Glomus tumors account for 1% to 5% of all soft-tissue tumors of the hand, but they can be found anywhere on the body. Extra-digital cases reported in the literature account for 12% to 65% of all glomus tumors. A review of 52 extra-digital tumors included locations on the thigh (n=11), calf/ankle (n=7), foot (n=4), buttocks (n=3), trunk (n=7), arm (n=9), and wrist/hand (n=7). Extracutaneous lesions have also been reported along the GI tract, on the lung, and in the urinary bladder. These lesions are much less common.

Glomus tumors should be in the differential for any violaceous, painful subcutaneous nodule. Other considerations should include blue nevi, blue rubber bllie nevus syndrome, Kaposi sarcoma, eccrine spiradenoma, leiomyoma, Merkel cell carcinoma, and venous malformation. The Love test and Hildreth sign are two useful findings to suggest the diagnosis of a glomus tumor. The Love test involves...
applying pressure to the nodule with a pencil tip or pinhead, causing an extreme, localized sensation of pain, and then placing a tourniquet proximal to the lesion to induce a temporary ischemia. In the Hildreth sign, the pain decreases upon application of the tourniquet. The Hildreth sign is 92% sensitive in diagnosing glomus tumors.10

If necessary, imaging of cutaneous tumors should begin with ultrasound. MRI can also be used but has proven less sensitive.9,10 The prognosis for solitary glomus tumors is excellent, and surgical excision is considered curative. If a glomus tumor recurs in the same location, it can be assumed that the primary tumor excision was incomplete. Recurrence rates in the literature vary from 0% to 33% after one year. Other curative treatments include injection of hypertonic saline, flash lamp tunable dye laser, and sclerotherapy.6,9 Malignant transformation is seen in only 1% of glomus tumors and is more likely in large tumors (> 2 cm), deep tumors, and those with high mitotic activity.2,7,9

Conclusion
The current case should serve as a reminder that although glomus tumors are commonly found on the hands, they can be found anywhere on the body. This type of tumor should be considered in the differential for any painful skin nodule, especially those demonstrating a violaceous tint. The Love test and Hildreth sign can provide clues to the diagnosis prior to removal and biopsy. Removal of the glomus tumor is generally curative and can relieve extreme pain almost instantly, limiting unnecessary testing and treatments.

References