Case Report

A Solitary Volar Extradigital Glomus Tumor Mimicking a Painful Ganglion

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Keywords
Glomus tumor · Extradigit · Ganglion

Abstract
Glomus tumors are neoplasms arising from the glomus body, which regulate skin temperature. They are mostly benign tumors and present in the subungual area of a distal phalanx. However, they can occur in extradigital location, and they may not be typical of intradigital glomus tumors. This makes it difficult to diagnose extradigital glomus tumors. We report a volar extradigital tumor mimicking a painful ganglion with a literature review.

Introduction
Glomus tumors are approximately 1–5% of all hand tumors [1]. They are kind of vascular neoplasms from glomus body which controls blood pressure and temperature by regulating blood flow in the cutaneous vasculature [2]. They have typical symptoms such as stabbing paroxysmal pain in the fingertips, tenderness, and cold intolerance [3]. Although these tumors could be found anywhere throughout the body, most of them occur in a distal phalanx of the fingers, often subungual area. If these tumors occur in extradigital site, it might be misdiagnosed such as ganglion, epidermal cyst, and neuroma. We present a case of extradigital glomus tumor mimicking a painful ganglion with a literature review.

Case Report
A 52-year-old female came to the outpatient clinic with a tiny palpable mass on her left hand. The mass, which had been growing for 10 months, induced a sharp stab of pain when touched. On physical examination, the mass was tiny, soft, and well-margined. The clinic which she visited first diagnosed a painful ganglion and recommended the surgery.

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Published by S. Karger AG, Basel

DOI: 10.1159/000511427
We performed a MRI with contrast. The MRI showed a well-defined rounded mass measuring 0.4 × 0.4 × 0.4 cm³ in the volar side of the third web space. The signal of mass was low-signal intensity on T1-weighted images and high-signal intensity on T2-weighted images. The mass revealed strongly homogeneous strong enhancement (Fig. 1). Therefore, the preoperative diagnosis would be an extradigital glomus tumor.

The patient underwent open excision of the mass under the brachial plexus block. The mass was located in the subcutaneous layer. The gross feature of the tumor was a yellowish, solitary, and well-capsulated tiny mass. The histological findings revealed the equally spaced cells, polygonal cells with identifiable cellular borders, moderate clear cytoplasm, and thin-walled blood vessels (Fig. 2). For the confirmation of glomus tumor, smooth muscle actin (SMA) and CD34 were stained in the tumor cells (Fig. 3). Symptoms improved immediately after surgery, and then there was no recurrence after the 1 year of follow-up.

**Discussion**

A glomus body is a neuromyoarterial apparatus located in the reticular dermis, which controls body temperature by regulating blood flow to the skin. A glomus tumor is a neoplasm of the normal glomus body. It was first described clinically by Wood [4] in 1812 and confirmed pathologically and named "glomus tumor" by Masson [5] in 1924.
Although glomus tumors could be occurred anywhere, most common site of occurrence is known to in the digits, especially subungual area [2]. Glomus tumors have been thought to be found much in extradigital area. This thought might result in delayed- or misdiagnosis about extradigital glomus tumors. According to TK Schiefer et al. [6], extradigital glomus tumors were found approximately 61% of 137 cases at their institution during a 20-year-period. Also, other study by Heys et al. [7] showed that 67% of 43 cases were extradigital tumors. Therefore, glomus tumors should be judged by not only location but also history, symptoms, and diagnostic images [6, 8].

Intradigital glomus tumor is common in female, while extradigital tumors are more common in men. Extradigital glomus tumors occur at a rate 4 times higher for men [6, 9]. These tumors are mainly diagnosed between the fourth and seventh decades.

Glomus tumors present classical symptom triad of pain, pinpoint tenderness, and hypersensitivity to cold [10]. However, these typical symptoms are often invisible, making it difficult to diagnose extradigital glomus tumors. A glomus tumor must be considered in the differential diagnosis of any soft tissue mass such as a ganglion, neuroma, hemangioma, angioma, and so on.

**Fig. 2.** Microscopic images revealed that equally spaced cells (cookie-cutter appearance), polygonal cells with identifiable cellular borders, moderate clear cytoplasm, and thin-walled blood vessels (H and E, ×200).

**Fig. 3.** Immunohistochemistry revealed diffuse, high SMA expression. SMA, smooth muscle actin.
The most frequent site of extradigital glomus tumors is forearm, and there are some case reports about forearm glomus tumors. Chronic pain with tenderness around forearm glomus tumors [11, 12], asymptomatic glomus tumor [13], and skin color change around tumors [14, 15] were reported. These case reports show that extradigital glomus tumors are very difficult to diagnose because of various symptoms.

MRI has been shown to be the most sensitive modality, detecting 82–90% of glomus tumors in the hand [16]. The typical appearance on MRI is a well-margined mass of intermediate or low signal on T1-weighted images and high signal on T2-weighted images with diffuse enhancement. The specificity of MRI for detecting glomus tumors is 50%, which is lower than high sensitivity [16].

A pathologic typical finding of glomus tumors is the regular cell spacing which is called “cookie-cutter appearance.” It looks like the cells were created with a cookie cutter because the spacing between cells is equal and they all look very similar.

The treatment of glomus tumors is a complete surgical excision. If total extirpation is not done due to unclear operation field by hemorrhage, the recurrence rate of glomus tumors is high [11]. Therefore, usage of tourniquet to operate without bleeding results in reduced recurrence rates.

In this case, the patient was a 52-year-old female. The patient’s gender was female though extradigital glomus tumor is developed in male dominantly. Of the triad of typical symptoms, only pain and pinpoint tenderness were shown except cold hypersensitivity. MRI result of the mass showed typical glomus tumor findings with enhancement pattern.

In conclusion, we presented an extradigital volar glomus tumor mimicking a painful ganglion. The diagnosis of extradigital glomus tumors is a challenge. Unusual location and nonspecific symptoms make the accurate diagnosis difficult. Hence, clinicians pay attention to history and diagnostic images besides symptoms.

Statement of Ethics

The patient provided written informed consent for the publication of clinical details and images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Funding Sources

The authors received no funding for this paper.

Author Contributions

D.G. Kim was involved in the surgery, data collection, manuscript preparation and its edition. S.R. Kang participated in the reviewing and interpretation.
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