

CASE REPORT

Diagnostic Dilemma of Subungual Glomus Tumor of Great Toe: A Case Report and Review of the Literature

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ABSTRACT

Background: Glomus tumor is a benign neoplasm also known as angioneuroma, arising from glomus bodies which are specialized structures for thermoregulation.¹ Solitary benign glomus tumors are small, usually <1.0 cm in size and they pose a diagnostic dilemma.^{2,3} They usually present with a classical triad of spontaneous pain, pressure tenderness, and cold hypersensitivity.

Case description: A 37-year-old woman presented to us with complaints of pain and temperature sensitivity, in left great toe with difficulty in wearing shoes from the last 5 years. X-rays and blood investigations were normal. Magnetic resonance imaging (MRI) was done and showed T1 hypointense, T2 hyperintensity, and subtle bone expansion with the possibility of glomus tumor. En Bloc removal of the lesion was done and a histopathological report confirmed it to be a glomus tumor. The patient was asymptomatic after 3 weeks of surgery.

Discussion: The purpose of this case report is to aware the orthopedic surgeon regarding the possibility of glomus tumor as a differential of chronic toe pain.

Keywords: Classical triad, En Bloc, Glomus tumor.

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INTRODUCTION

Glomus tumor is a benign neoplasm, arising from neuromyoarterial apparatus called glomus bodies which are specialized structures for thermoregulation.⁴ Glomus bodies are arteriovenous shunts surrounded by glomus cells, which contract via sympathetic activity, to regulate blood flow to the extremities. Glomus bodies are located in the dermis but more densely in apical skin areas, primarily the fingertips, followed by the base of the foot and the rest of the body. Glomus tumor has been reported at body sites, such as, head, neck, colon, lung, tongue, stomach, ear, elbow, wrist, hand, foot, toes, bladder, patella, coccyx, rectum, penis, and cervix.⁵ Glomus tumors are most commonly found in the extremities, with up to 75% of the presentations diagnosed in the hand and 90% of the hand lesions are located in the subungual tissues of the fingers.⁶

Foot glomus tumors are relatively rare due to the lower concentration of glomus bodies in the foot (i.e., even in the subungual region of the toes).⁷ Due to the low incidence of foot glomus tumors and variable clinical presentations, delayed diagnosis is frequent.⁸⁻¹⁰ In the foot, a clinical presentation has been reportedly confused with Morton's neuroma, flexor hallucis longus tendon injury, plexiform neurofibroma, and an ingrown toenail.¹¹⁻¹⁴

Glomus tumor occurrences have been reported mostly in females during the third through fifth decades of life, though, the lesion may occur at any age. A glomus tumor most commonly presents as a small solitary blue or purple nodular subungual lesion clinically.¹⁵ Patients usually present with the classical triad of spontaneous pain, pressure tenderness, and hypersensitivity to cold.¹⁶ Majority of the lesions are solitary but multifocal involvement can occur and is seen in cases of neurofibromatosis type I. Sometimes the history of trauma in past may be present.

Despite advances in imaging capabilities, histopathological examination of the excised tumor is absolutely necessary to provide a definitive diagnosis.² Microscopically, the lesion shows uniform epithelial cells distributed outside a branching vasculature with pink cytoplasm and centrally placed nuclei (Figs 1 to 8).

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Treatment is mainly surgically requiring complete excision of the lesion. Despite the relative rarity of this entity, one must be aware of the possibility of glomus tumor when a patient presents with persistent pain around his or her nail bed and has experienced previous unsuccessful surgery for pain.¹⁷ We believe that reporting this case may help to fill lacunae in clinical knowledge related to glomus tumors and help orthopedicians to keep this in mind while dealing with such cases.

CASE DESCRIPTION

A 37-year-old woman presented in the outpatient department with complaints of pain and temperature sensitivity, in her left great toe with difficulty in wearing shoes for the last 5 years. The pain was acute in onset, spontaneous, paroxysmal, sharp, and worsened upon exposure to cold weather. The patient had consulted a dermatologist and a general surgeon and was managed with analgesics and partial nail plate removal and antibiotics but had no improvement. There was no history of trauma, similar swelling elsewhere, fever, diabetes mellitus, hypertension, and pulmonary tuberculosis.



Fig. 1: Preoperative



Fig. 2: Intraoperative



Fig. 3: Intraoperative



Fig. 4: Biopsy sample



Fig. 5: Postoperative

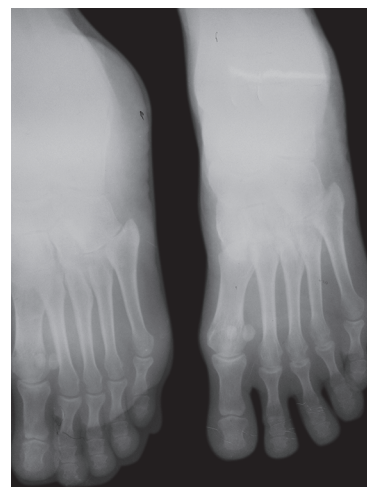


Fig. 6: X-ray foot-anteroposterior and oblique view

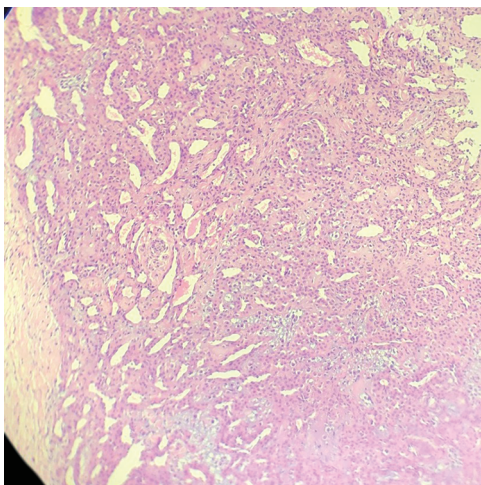


Fig. 7: H and E stained tissue showing glomus cells

Examination revealed subungual swelling of 0.8×1 cm over left great toe. Severe point tenderness was present directly over the nail plate with partial cicatrization of the nail plate. There were no trophic changes or sinuses seen. A provisional diagnosis of glomus tumor was made.

The plain radiograph showed no features of soft tissue or bony involvement. Haemoglobin (Hb) was 11.2 g/dL, white blood cell (WBC) was 6,500 μ L, erythrocyte sedimentation rate (ESR) was 12 mm/hour, c-reactive protein (CRP) was negative, fasting blood sugar was 70 mg/dL were done and found to be within normal limits. Magnetic resonance imaging (MRI) was done and showed T1 hypointense, T2 hyperintensity, and subtle bone expansion with the possibility of glomus tumor.

Written and informed consent was taken, and the patient was prepared for removal of the lesion under regional anesthesia (ring block).

The lesion was excised En Bloc after removing the nail plate. Grossly lesion was 0.8×1 cm in size, pale pink, well-capsulated, soft, and fleshy without signs of invasion to surrounding tissues. Removed tissue was sent for histopathological examination and diagnosis was confirmed on the report. The patient had disappearance of her symptoms by a week and became asymptomatic after 3 weeks of surgery without any complications. She has been under regular follow-up in the orthopedics outpatient department for 1 year.

DISCUSSION AND REVIEW OF LITERATURE

Glomus tumor is a benign neoplasm arising from neuromyoarterial apparatus called a glomus body. Glomus bodies are arteriovenous shunts, surrounded by glomus cells, which contract to regulate blood flow to the extremities. Hence, they play a role in thermoregulation.

The most common site of occurrence of this tumor is the subungual area of the fingers and sometimes in the toes. The glomus tumor was first mentioned by Wood in 1812¹⁸ but a complete description of its character was given by Masson in 1924.¹⁹ It accounts for <2% of soft tissue tumors.²⁰ The initiating, or causative, event associated with glomus cell proliferation and eventual glomus tumor formation is unknown. Once established, glomus tumors are typically composed of three components: glomus cells, vessels, and smooth muscle cells. According to the proportion of these three components, glomus tumors can

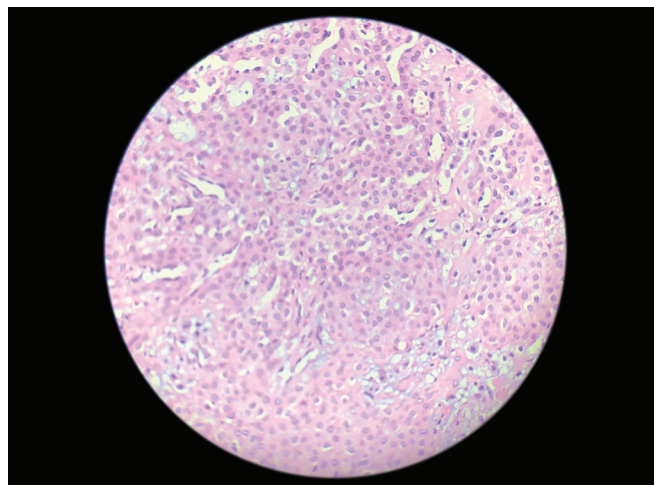


Fig. 8: Well-circumscribed lesion consisting of sheets and trabeculae of glomus cell surrounded by branching capillary sized as well as dilated blood vessels in a myxoid stroma. The cells are round with eosinophilic cytoplasm and round, sharply punched out nucleus with homogeneous chromatin and inconspicuous nucleoli

be subcategorized as solid glomus tumors, glomangioma, or glomangiomyoma. Solid glomus tumor is the most common variant (75%), followed by glomangioma (20%) and glomangiomyoma (5%).²⁰ The diagnosis is essentially clinical which is usually confirmed on histopathology with surgical excision.

This case showed that glomus tumors are very painful tumors and are challenging to diagnose and often the patient is misdiagnosed between different departments. Clinical examinations may include the Hildreth test and the Love test. The Hildreth test is highly specific and sensitive due to the vascular nature of the lesion. A positive Love test consists of pain on pressure directly over the symptomatic lesion.¹⁰ Differential diagnoses include ganglion cyst, hiradenomas, neuroma, angioma, melanoma, myxoid cyst,² melanoma, chronic paronychia, gout arthritis, and foreign-body granuloma.

Imaging modalities used in the diagnosis of a glomus cell tumor include plain film radiography, color Doppler ultrasonography, computed tomography, angiography, and MRI.²¹

Some patients give a history of preceding trauma for which they are managed for subungual hematoma and some are taking analgesic and antidepressant medications for chronic paronychia for many years. However, with proper history, examination, and investigations (MRI), the tumor can be diagnosed early and timely managed.

About 75% of glomus tumors will arise in the hands, with 75–90% of these in subungual locations.¹⁶ Glomus tumors have been categorized into solitary and multiple types. Most have been solitary, with up to 25% of the multiple types.^{22,23} Solitary glomus tumors will be encapsulated, will usually be found in subungual locations, and will contain numerous small vascular lamina.²⁴ In contrast, multiple type tumors will be unencapsulated, will rarely be subungual, will have larger shaped vascular spaces, and will often be asymptomatic.²⁵ Glomus tumors have been more common in females than in males and have been the most prevalent during the fourth decade of life.^{16,26}

About 1% of all glomus tumors are malignant.¹¹ The malignant histopathological features include large size (2 cm) and deep location, or moderate-to-high nuclear grade and increased mitotic

Table 1: Summarized descriptions of glomus tumors of toes given by different authors

S. no.	Author/year	Age/sex	Presentation	Diagnostic issues	Confirmation of diagnosis	Management
1.	Koti et al. ³³ 2001	31/F	Intermittent pain over the base of the nail bed of the right hallux	Several year histories	MRI, histopathological	Excision
2.	Pater and Marks ¹² 2004	28/F	Lateral nail pain over left hallux	Ingrown toenail	MRI, histopathological	Excision
3.	Seo et al. ³⁴ 2014	30/F	Pain in her right hallux	Inflammation or edema of the nail bed	MRI, histopathological	Excision
4.	Lui and Mak ¹⁰ 2014	41/F	Tiny lump and pain at her left great toe pulp	Neuroma, hemangioma	MRI, histopathological	Excision
5.	Mohindra et al. ¹ 2016	19/F	Bursting pain in the left great toe	Pain since past 8 years	MRI, histopathological	Excision
6.	Current report	37/F	Bursting pain and temperature sensitivity in left great toe with difficulty in wearing shoes	Chronic paronychia, ingrown toe from last 5 years	MRI, histopathological	Excision

rate (5/50 high power fields), or the presence of atypical mitotic figures. If the histological criteria for malignancy are met, then the lesions are associated with more than a 25% risk of metastasis.²⁷ However, tumors with some but not all of these ominous features (i.e., high mitotic activity and superficial location, large size only, or deep location only) are best described as glomus tumors with uncertain malignant potential, or symplastic glomus tumors (high nuclear grade only).²⁸

A differential diagnosis should be carried out that considers the following possibilities: chronic paronychia, where a complete and detailed clinical history of the patient can serve to easily determine if this diagnosis is, in fact, the case; subungual melanoblastoma, which, unlike glomus tumors, is not usually painful; subungual hematoma, which is usually accompanied by a previous trauma and as a result could have been forgotten about; and other less frequent possibilities, such as, neuron, plexiform neurofibroma, papillomas, and fibromas. Last but not least, Koenen's tumors should also be taken into account since they appear as medial or lateral fleshy elongated proliferations that are located on the nail and closely joined to the matrix.^{29,30}

Carroll and Berman¹⁶ have defined the classic triad of symptoms of glomus tumors as severe pain, point tenderness, and cold sensitivity, and these symptoms form the key to the diagnosis. However, not all glomus tumors will present with this classic triad of symptoms.

Van Geertruyden et al.³¹ have reported that in patients with solid glomus tumors, pain, point tenderness, and cold sensitivity will occur at a rate of 80, 100, and 63%, respectively.

In Table 1, we have summarized descriptions of glomus tumors of toes given by different authors.

In conclusion, the diagnosis of foot glomus tumors requires clinical suspicion given its low incidence and wide differential diagnosis. Patients may have undergone prior surgeries as a result of misdiagnosis (most frequently an ingrown toenail).³²

We believe that reporting this case may help to fill lacunae in general clinical knowledge related to glomus tumors and help clinicians. En Bloc excision of the tumor is a good modality of treatment. There is generally a delay in diagnosis and often the patient suffers great morbidity and having knowledge of this can help to reduce it.

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