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ORIGINAL RESEARCH

Understanding Nail Glomus Tumor: History, Pathology, and Treatment

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ABSTRACT
Glomus tumors are uncommon benign vascular neoplasms frequently located in the hand, particularly in the subungual region. Despite their unclear etiology, various hypotheses attempt to explain their pathogenesis and the associated pain mechanism. These tumors typically manifest as a bluish or pinkish-red discoloration of the nail plate, accompanied by a classic triad of localized tenderness, severe pain, and sensitivity to cold. Differential diagnosis should consider other painful finger lesions such as leiomyoma, eccrine spiradenoma, haemangioma, neuroma, osteochondroma, or mucous cyst. Diagnostic evaluation includes clinical tests like Love's pin test, Hildreth's test, and transillumination, complemented by imaging modalities such as magnetic resonance imaging (MRI), ultrasonography, and radiography. Surgical excision is imperative for complete symptom resolution and to prevent recurrence. Various surgical approaches are documented, chosen based on tumor location and individual surgeon preference.The cases described are instances of glomus tumors, which are rare benign neoplasms originating from the glomus body, a specialized arteriovenous structure involved in thermoregulation.Case studies describe patients with longstanding thumb pain and nail changes, cold-sensitive pain in the left little finger, recurrent pain in the left middle finger, pregnant woman with severe episodic pain in the right index finger during pregnancy and pain under the left second toenail. Patients were diagnosed via MRI and leading to surgical excision of a glomus tumor confirmed on histopathology. This article reviews key aspects of glomus tumors in the hand, encompassing their etiology, clinical presentation, diagnosis, management strategies, and recurrence prevention.

INTRODUCTION

Nail glomus tumors are rare benign neoplasms that typically arise from the glomus body, a specialized structure involved in temperature regulation in the dermis. These tumors are characterized by their painful nature and are often found in subungual regions, making them a significant concern in dermatology and hand surgery [1]. Glomus tumors are uncommon, benign vascular growths that originate from the glomus body, a contractile neuromyoarterial structure located within the reticular dermis [2]. The glomus body comprises an afferent arteriole, an anastomotic vessel called Sucquet-Hoyer canal, a primary collecting vein, intraglomerular reticulum, and a capsular portion. This structure plays a role in regulating blood pressure and temperature by controlling blood flow in the skin's vasculature. Hyperplasia within any part of the glomus body can lead to the formation of a tumor [3].

While glomus tumors can occur anywhere on the body, they most commonly appear on the distal phalanx of the fingers, especially in the subungual region, particularly among females [4]. In contrast, males tend to develop these tumors in other body areas. There are generally two types of glomus tumors: solitary and multiple. Solitary glomus tumors are more prevalent, whereas multiple glomus tumors rarely affect the fingers [5]. Multiple glomus tumors can sometimes occur concurrently with type 1 neurofibromatosis and are often asymptomatic, posing a diagnostic challenge. Magnetic resonance imaging (MRI) is highly effective for detecting glomus tumors and detailing their anatomical characteristics, including size and location [6]. Complete surgical removal of the tumor is typically recommended to minimize the risk of recurrence [7, 8]. This article explores the historical milestones, pathology, clinical presentation, diagnosis, and treatment options for nail glomus tumors.

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HISTORICAL MILESTONES

The history of nail glomus tumors dates back to the early 20th century when they were first described by Hildreth in 1924. He recognized them as a distinct entity and noted their tendency to cause severe pain disproportionate to their small size. Throughout the 20th century, several key studies further elucidated the pathophysiology and clinical characteristics of glomus tumors. Notably, in 1972, Smith and Barnard published a seminal paper detailing the histological features of glomus tumors, which confirmed their origin from the glomus body.

ETIOPATHOGENESIS

The exact cause of glomus tumors remains uncertain and may be influenced by factors such as gender, age, trauma, or genetic predisposition. Some researchers hypothesize that trauma could lead to reactive hypertrophy in the glomus body due to underlying structural weaknesses. Recently, studies have identified a familial variant of glomus tumor associated with chromosome 1p21–22, involving truncating mutations in the glomulin gene. This gene encodes a 68-kDa protein of unknown function [9, 10].

The mechanism of pain in glomus tumors is not fully understood, but several theories have been proposed. It is suggested that the presence of a sensitive capsule, which responds to pressure, and mast cells releasing substances like heparin, histamine, and 5-hydroxytryptamine may sensitize pressure and thermal receptors. Additionally, the dense innervations of non-myelinated nerve fibers within glomus tumors have been proposed as a potential cause of pain [11].

Glomus tumors originate from the glomus body, which is a specialized arteriovenous structure involved in thermoregulation. These tumors are typically small, measuring less than 1 cm in diameter, and are found predominantly in the subungual region of fingers. Histologically, they are composed of glomus cells, which are modified smooth muscle cells surrounded by an extensive vascular network. Immunohistochemical studies have shown that glomus tumors express markers such as smooth muscle actin and type IV collagen, confirming their smooth muscle origin [12].

CLINICAL PRESENTATION

The hallmark symptom of a nail glomus tumor is severe paroxysmal pain exacerbated by cold temperatures or pressure. Patients often describe the pain as sharp, localized, and intolerable. The pain may worsen at night, disrupting sleep and daily activities [13]. Physical examination typically reveals a small bluish or reddish nodule under the nail bed. Due to their small size and location, these tumors can be challenging to diagnose clinically and may require imaging studies for confirmation.Glomus tumors constitute 1–5% of soft tissue tumors affecting the hand, with approximately 75% of cases located under the nail (subungual). Less frequently, these tumors occur in other areas of the hand such as the nail matrix, nail bed, and fingertip pulp. They predominantly affect middle-aged women [14].

Typically, glomus tumors appear as small, slightly raised nodules that can be bluish or pinkish-red in color. They are often painful and can cause elevation, deformation, and discoloration of the nail when located beneath it. The classic clinical presentation includes localized tenderness, severe pain, and sensitivity to cold, forming a distinctive triad suggestive of a glomus tumor. Patients frequently report worsening symptoms in cold weather, when handling cold objects, or after exposure to cold water [15, 16].

DIAGNOSIS

Diagnosis of nail glomus tumors is primarily clinical, based on history and physical examination findings. Radiographic imaging, such as ultrasound or magnetic resonance imaging (MRI), can aid in localizing the tumor and assessing its size and relationship to adjacent structures. MRI, in particular, is sensitive for detecting small glomus tumors and can provide valuable information for surgical planning.

In addition to the classic clinical presentation, there are three valuable diagnostic tests for identifying glomus tumors. The Love's pin test involves applying pressure with a pinhead to the suspected area, eliciting severe pain if a glomus tumor is present. Hildreth's test utilizes a tourniquet on the arm to induce temporary ischemia; if the patient experiences relief from pain in the affected area, it suggests the tumor's vascular nature. This relief can be confirmed by repeating the Love's pin test painlessly under the inflated tourniquet, with pain returning upon tourniquet removal. The cold-sensitivity test involves applying cold water or an ice cube, causing increased pain in patients with glomus tumors [16].

Another less commonly used test is transillumination, where light is passed through the finger pad to reveal a red opaque image, showing sensitivity of 23–38% and specificity of 90%. Netscher et al. [17] found the cold-sensitivity test to have 100% sensitivity, specificity, and accuracy, while Love's pin test showed 100% sensitivity and 78% accuracy. Hildreth's test was most specific, with 100% specificity, 71.4% sensitivity, and 78% accuracy [18].

Differential diagnosis should consider other painful tumors like leiomyoma or eccrine spiradenoma. Additionally, conditions such as hemangioma, neuroma, or gouty arthritis can mimic glomus tumors, posing diagnostic challenges. Multiple glomus tumors need differentiation from cavernous hemangioma and blue rubber-bleb nevus syndrome due to potential confusion [19].

Despite classic symptoms, diagnosing glomus tumors promptly remains challenging due to variable symptom presentations, including chronic pain and

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hypersensitivity. Misdiagnosis as neuropathic complaints, arthritis, or neuralgia can lead to inappropriate treatments like sympathetic ganglionectomy or radicotomy. Furthermore, the rarity and difficulty in detecting impalpable glomus tumors during examination contribute to delays in accurate diagnosis [20].

TREATMENT OPTIONS

Surgical excision remains the treatment of choice for nail glomus tumors. The goal of surgery is complete removal of the tumor while preserving nail function and aesthetics. Advances in surgical techniques, including nail-sparing approaches, have improved outcomes and reduced postoperative complications such as nail deformity. In cases where surgery is contraindicated or in patients who are not surgical candidates, alternative therapies such as cryotherapy or sclerotherapy may be considered, although their efficacy compared to surgery is debated [21-23].

FACTORS AFFECTING RECURRENCE

The literature reports a recurrence rate of approximately 4–50% following surgical excision of glomus tumors [24]. Early recurrence is typically attributed to incomplete excision or the presence of an undiagnosed second tumor at the initial operation. In contrast, "late recurrence" involves the development of a new lesion at or near the site of the previous excision. Subungual tumors tend to have a higher probability of recurrence, possibly due to the surgical approach focusing on conservatively excising matrix tissue to prevent postoperative nail plate deformities [25].

The approach of making larger incisions can lead to complications such as prominent postoperative scars, paresthesias from greater nerve branch injury, and nail dystrophy. Skin-colored tumors, unlike the classic red, blue, or purple glomus tumors, present challenges during surgery due to difficulty in clear delineation and proper margin identification, potentially leading to incomplete excision and increased recurrence risk [26].

Studies by Foucher et al. [27] indicated a 7% recurrence rate after three to five years, while Heim and Hanggi's series showed a nearly equal distribution between early and delayed recurrences, with a slight majority in early recurrences (54%). Clinically, persistent symptoms of glomus tumors lasting more than three months should prompt re-exploration of the affected area and repeat imaging to assess for recurrence.

Case 1: A 26 year old female was referred from the surgery department with a history of left thumb pain persisting for 5 years. Initially mild, the pain intensified over time, exacerbated during rainy and winter seasons or following minor trauma. Three years ago, she noticed her nail splitting, which progressively involved the entire nail length. Her medical history was otherwise unremarkable. On examination, a tender swelling with ill-defined margins was found behind the proximal nail fold. The nail exhibited a longitudinal split extending from the free end to the proximal fold. No discoloration was observed in the nail or proximal fold. Love's test indicated sensitivity, while Hildreth's test was negative. Cold water immersion induced pain, suggesting cold sensitivity. Routine blood and urine tests were normal, and radiographic findings were unremarkable (Figure 1a).

Surgical exploration under digital nerve block and tourniquet revealed a semitranslucent mass measuring 2 mm × 3 mm, which was excised and sent for histopathological examination. Histopathology showed clusters of uniform tumor cells within a hyalinized stroma. These cells displayed sharply punched-out nuclei and distinct borders. Dilated vessels containing tumor cell clusters were also identified within their walls (Figure 1 b and c).

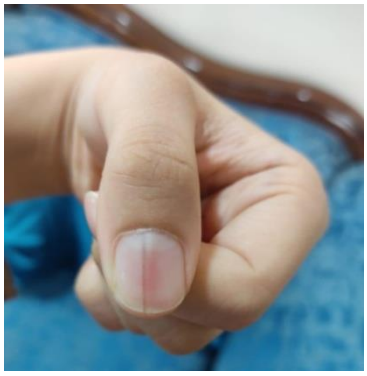

Figure 1a: Fullness in nail fold region

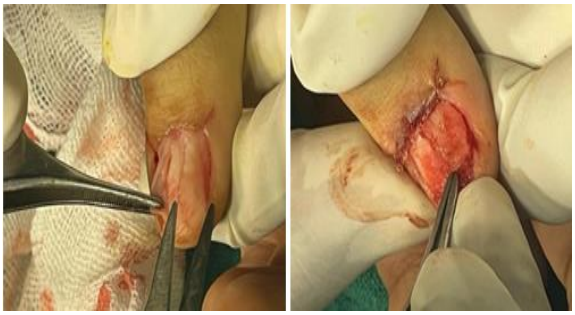

Figure 1b and c: Glomus tumor during exploration

Case 2: A 39 years old female presented to outpatient department with a five years history of pain in left little finger nail. No history of any preceding trauma. The nail is slightly discolored. She described the incidence of severe pain when exposed to cold. There was no evidence of Raynaud's phenomenon. There was absence of fever, rash or ulcer.

On examination, a sharp localised point of tenderness was found over the nail of the left little finger. Love's pin test and Hildreth's test was positive. Cold sensitivity test was equivocal. There was a slight discoloration nail. There was no rise of local temperature, regional lymph node enlargement and

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the patient was afebrile. The systemic examination revealed normal.

MRI FINDINGS

There isointense (to muscle) subcutaneous lobulated soft tissue lesions (0.7 × 0.6 cm) on T1W1, hyperintense on T2W1 and STIR image were seen at dorsal aspect of distal phalanx of left little finger. The lesions exhibited mild heterogeneous enhancement on post contrast T1W1. Thin curvilinear vessels with flow-void in all sequences were seen within the vicinity of the lesions. All these features were suggested of glomus tumor at dorsal aspect of distal phalanx of left little finger.

SURGICAL PROCEDURE

The procedure was carried out under digital block with 1% xylocaine and proximal tourniquet control. A surgical marker was used to mark the location of the tumor on the nail plate. A direct transungual approach was used and proximal nail avulsion was performed to expose the nail bed. A longitudinal incision was made on the nail bed over the tumor. A 0.7 × 0.6 cm shiny, pinkish, encapsulated lesion emerging from the nail bed was discovered. The tumor and its fibrous capsule were carefully dissected. Then we repaired the nail bed. The excised tumor was sent to histopathology, which confirmed the diagnosis.

Case 3: A 31 year old female presented with a painful swelling on the distal digit of her left middle finger that had been bothering her for 12 years. The swelling was particularly painful when exposed to cold, pressure, or accidental trauma. She had undergone surgery previously for the same issue, but the swelling and pain returned within 6 months.

Upon dermatological examination, the terminal digit of her left middle finger appeared bulbous with a deformed, slightly bluish nail plate and extreme tenderness over the nail. Hildreth's sign and Love's test were positive, indicating sensitivity, but cold sensitivity could not be elicited. There was noticeable wasting of the left hand and forearm due to disuse. X-rays did not reveal any abnormalities.

Surgical exploration through a transungual approach revealed a shiny, encapsulated tumor, which was completely excised. Biopsy of the tumor confirmed it to be a glomus tumor.

Case 4: A 35 year old pregnant woman who is pregnant presented with an 11 year history of severe, intermittent shooting pain in the tip of her right index finger. The pain lasted only a few seconds each time but was extremely intense and had become more frequent during her pregnancy. Touching the finger or accidental injury triggered intense pain. Elevating the limb provided partial relief. On examination, there was a slight bulbous swelling at the fingertip, but immersing the finger in ice-cold water did not provoke pain. X-rays and high-resolution CT scans showed no abnormalities.

During surgery using a transungual approach, a pearly, shiny, encapsulated swelling measuring 2 mm in diameter was identified and removed with a 3 mm margin. Histopathological examination confirmed the clinical suspicion and intraoperative diagnosis of a glomus tumor.

Case 5: A 43-year-old woman presented for consultation, having undergone cholecystectomy 26 days earlier. She complained of pain at the tip of her right middle finger persisting for 6 months, with no history of preceding trauma. The pain worsened upon exposure to cold and minor injuries. Changes in her nail's color and shape marked the onset of symptoms, characterized by a single-finger pseudo-clubbing of the nail on the right middle finger with anterior curvature, V-shaped anterior onycholysis with greenish discoloration, and a visible erythronychic band. Dermoscopic examination revealed lunula effacement and enhanced visualization of the V-shaped onycholysis, resembling a tree trunk pattern at the level of the erythronychic band. Hand X-ray showed a well-defined soft tissue lesion on the dorsal aspect of the distal phalanx of the third right finger, along with bone deformation but no osteolysis.

Surgical excision of the tumor was performed, with intraoperative dermoscopy of the nail bed revealing a purple-red area and regular linear vessels. The tumor was enucleated, and Dubois technique surgery was performed to reduce the anterior edge. Histopathological examination revealed a proliferation of tumor cells arranged around numerous vascular spaces lined by flattened endothelial cells. These vessels had thickened walls and were surrounded by layers of ovoid cells with regular, round nuclei and scant eosinophilic cytoplasm [Figure 5]. Postoperatively, the nail grew normally over the next 3 years without abnormalities of the nail plate or functional issues.

Case 6: The patient, a 52-year-old female, reported experiencing pain beneath her left second toenail for about 15 months. On physical examination, she exhibited severe pain upon palpation of the left second toenail. Discoloration was observed beneath the nail, accompanied by slight separation of the nail from the nail bed at the proximal end, which overlaid the discolored area and soft tissue mass. An MRI was performed, revealing a 6 × 3 mm high signal intensity mass located in the subungual region of the left second digit. There were no signs of periosteal reaction, soft tissue edema, or abnormal bone marrow signal. These findings were indicative of a glomus tumor, with an epidermal inclusion cyst considered as a differential diagnosis.

The decision was made to remove the nail and perform surgical excision. In the operating room, the nail was avulsed, and the nail bed was carefully examined. The tumor was found to extend from the center of the nail bed proximally toward the nail matrix and deep under the eponychium. A longitudinal incision was made along the nail bed, and

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dissection revealed a well-encapsulated soft tissue mass. The mass, measuring approximately 7 mm in diameter, was completely removed with meticulous attention to ensure no residual tissue remained. The surgical site was irrigated, and the nail bed was repaired using 4-0 Monocryl sutures in a simple interrupted technique.

CONCLUSION

Nail glomus tumors are rare but clinically significant lesions characterized by intense pain and localized under the nail bed. Understanding their historical context, pathological basis, clinical presentation, and treatment options is crucial for dermatologists, hand surgeons, and primary care providers. Advances in imaging techniques and surgical approaches continue to refine the management of these tumors, aiming to improve patient outcomes and quality of life. Glomus tumors typically present with severe pain, exacerbated by cold or pressure, and often involve changes in the nail or surrounding tissue. Diagnosis relies on clinical findings, imaging (MRI), and histopathology. Treatment involves surgical excision, often through a transungual approach for subungual tumors. Prognosis is excellent following complete excision, with resolution of symptoms and low recurrence rates. Early recognition and management are crucial to alleviate symptoms and prevent complications from chronic pain and disability. Complete surgical excision is essential for achieving symptom relief and minimizing the risk of recurrence. Adequate excision can be ensured through comprehensive preoperative assessment, including thorough physical examination and appropriate imaging studies, coupled with meticulous surgical technique.Various surgical approaches are documented in the literature, with surgeons often choosing techniques based on the specific anatomical location of the tumor and their individual preferences.

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