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(CASE REPORT)

Glomus tumor of the great toe: An uncommon case report

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Abstract

Introduction: Masson's glomus tumor is a neuromyoarterial hamartomatous proliferation that develops from corpuscules located at the dermal-hypodermal junction, it is a rare but not exceptional benign tumor and it is less frequently reported in the foot.

Case presentation: We present a unique case based on its unusual location, clinical presentation (pain being the primary symptom) and its misleading appearance on MRI. Treatment involved surgical excision of the tumor, resulting in a favorable outcome. Histopathological examination of the excised specimen remains crucial for definitive diagnosis, highlighting the importance of considering this unusual location in the differential diagnosis.

Discussion: Pathogenesis remains poorly understood, and diagnosis can be difficult. It preferentially affects the hands (75%) and it is subungual in 50% of cases. Diagnosis is based on the clinical triad of sensitivity, pain and intolerance. Advances in high-resolution MRI have improved false negatives, with lesions showing hypo-signal in T1 and hypersignal in T2, with clear contrast between normal tissue and tumor. Diagnostic certainty is provided by histology. After complete removal of the tumor, results are generally considered excellent. In most cases, pain disappears rapidly, and the affected finger or limb returns to normal within three months.

Conclusion: Glomus tumors are certainly rare, but not exceptional, and a high degree of suspicion is required when faced with a painful purplish nodule. Usually distal, periungual in location, they can occur anywhere, with sometimes unusual and puzzling localizations.

Keywords: Glomus tumor; Pain; Hallux; Vascular

1. Introduction

Glomus tumors are uncommon benign neoplasms arising from the thermoregulatory glomus body, a specialized microvascular system. They are most frequently located in the subungual tissue[1]. It mainly affect the digital extremities, but extra-digital localizations are not uncommon and are often unrecognized, leading to delays in diagnosis and management [2]. Histopathological examination of the excised specimen remains crucial for definitive diagnosis.

We report a case of glomus tumor with an atypical presentation and location, which was definitively treated with surgical excision with good evolution.

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2. Case presentation

This is a 50-year-old female patient, with no particular pathological history, who has presented for 2 years with intermittent pain located at the right big toe. We saw the patient, who consulted us several times in view of the increasing intensity of her pain and the lack of improvement with symptomatic treatment.

Clinical examination of this patient showed a painful nail, which is exacerbated on palpation with positive love's sign. Osteoarticular examination of the right hallux was unremarkable, and standard radiographs and laboratory tests were normal. MRI showed a subungual tumor of the great toe measuring 12 mm long axis, initially suggesting a glomus tumor.

Direct trans-ungual approach was performed, then, the tumor was excised, and pathological examination confirmed the diagnosis of a glomus tumor. The postoperative course was straightforward, with immediate pain relief, and the patient's clinical examination after 2 years showed no recurrence of symptoms

3. Discussion

A glomus tumor is a non-cancerous growth cells that develops from neuromyoarterial apparatus called a glomus body. These glomus bodies act like valves that control blood flow in the fingers and toes, helping to keep them warm [3].

Glomus tumors have been widely reported in a number of anatomical locations, including the knee, liver, trachea, auricle, lung, stomach, and fingers of the hand. Glomus tumors account for 1% to 5% of all hand tumors. Approximately 75% of reported glomus tumors have localized to the hand, and around 65% of these have occurred in subungual tissues due to the high concentration of glomus bodies in the subungual area. Intraosseous areas (interosseous glomus tumor) with glomus tumors have also been reported, notably in the ankle, proximal ulna, distal phalanx of the toes or fingers, and the fibula [4].

These tumors have been infrequently reported to occur in the toes, either subungually or interosseously. To our knowledge, finger pulp is the only reported location for this tumor. There are no documented cases in the toe pulp. Their rarity in the toes often presents a diagnostic challenge, leading to delays in diagnosis. The diagnosis of this lesion relies primarily on clinical examination, and surgery is the only definitive treatment option [5].

Clinically, the classic triad of pain, point tenderness upon palpation, and hypersensitivity to cold temperatures is a hallmark feature of glomus tumors [1] :

Spontaneous paroxysmal pain is rare at onset, but becomes habitual after a long evolution. It is reported in the majority of series. Its searing, excruciating nature contrasts with the discretion or absence of local signs.

Excruciating pain triggered by minimal pressure or the slightest contact on the affected area. This sign is pathognomonic and frequently reported in literature.

Increased sensitivity to pain triggered by changes in temperature, particularly cold, this feature is reported by the majority of authors.

Clinical examinations may include the Hildreth test and the Love test. The Hildreth test, due to the vascular nature of the lesion, boasts high specificity and sensitivity. A positive Love test indicates localized pain upon direct pressure over the symptomatic area. Differential diagnosis encompass ganglion cyst, hiradenomas, neuroma, angioma, melanoma, myxoid cyst, chronic paronychia, gout arthritis, and foreign-body granuloma [3].

There are no specific imaging techniques for confirming the diagnosis, but ultrasound, despite its low specificity, helps to localize the lesion. MRI remains the gold standard for the diagnosis of glomus tumors, as it enables precise localization of the lesion and its relationship with neighboring structures [2]. However, given the small size of these lesions, a targeting examination is crucial for proper diagnosis. Therefore, a high degree of clinical suspicion is necessary before resorting to MRI, which shows these tumors as well-defined masses with low signal on T1-weighted images and a high signal on T2-weighted images. Glomus tumors typically show intense enhancement on MRI after gadolinium administration due to their partly vascular nature. However, this MRI finding lack specificity for glomus tumors and can also be seen in other well-defined solid tumors, such as hemangiomas, venous malformations, neurilemmomas (Schwannomas), traumatic neuromas, mucoid cysts, melanoblastoma, plexiform neurofibromas, papillomas, fibromas,

Koenen's tumor, and blue rubber bleb nevus syndrome (which presents with soft, bluish nodules). In cases with atypical presentation, a skin biopsy might be necessary for a definitive diagnosis [4].

Glomus tumors have been extensively studied in terms of their pathological features. Typically, the gross specimen appears as well-encapsulated, grey-pink lesion. Microscopically, it resembles a normal neuromyoarterial arcade (glomus body) with a marked hypertrophied elements. This glomus body is composed of enlarged glomus cells nestled amongst abundant vascular channels. Notably, these cells contain relatively small concentrations of nerve fibers, despite the excruciating pain they can provoke. Fortunately, these lesions are almost always encapsulated, exhibiting minimal local invasion and no documented cases of metastasis [1].

Surgical excision is the mainstay of treatment, leading to prompt pain relief. Recurrence is uncommon but can occur, typically due to incomplete removal of the tumor. To minimize this risk, some authors advocate for resection margins exceeding the visible tumor borders. Complete surgical excision typically leads to complete resolution of symptoms. This was certainly the case for our patient, who returned to full function without pain [2, 6].

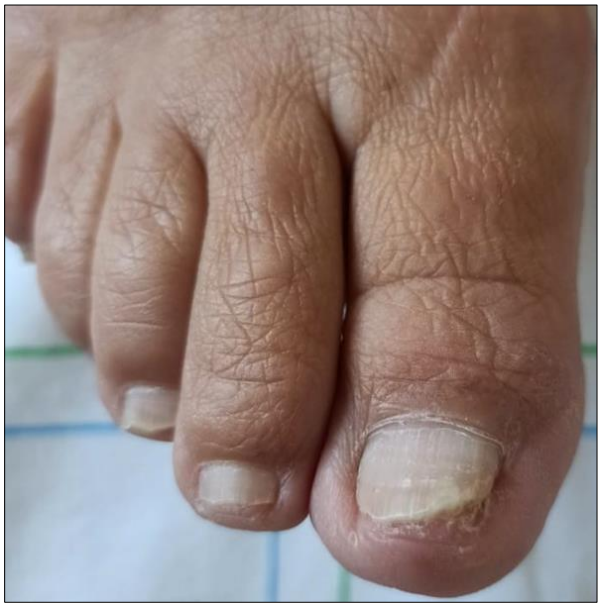

Figure 1 Normal clinical aspect of the great toe

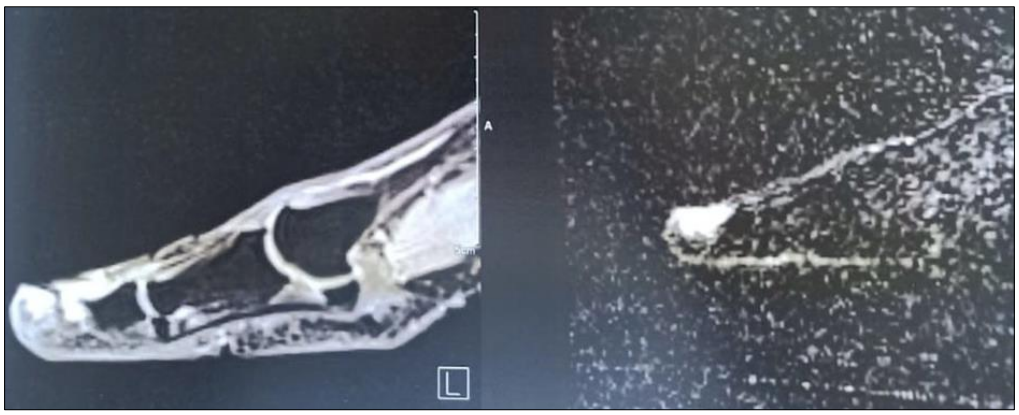

Figure 2 MRI (T2) shows subungual tumor of the great toe measuring 12 mm long axis

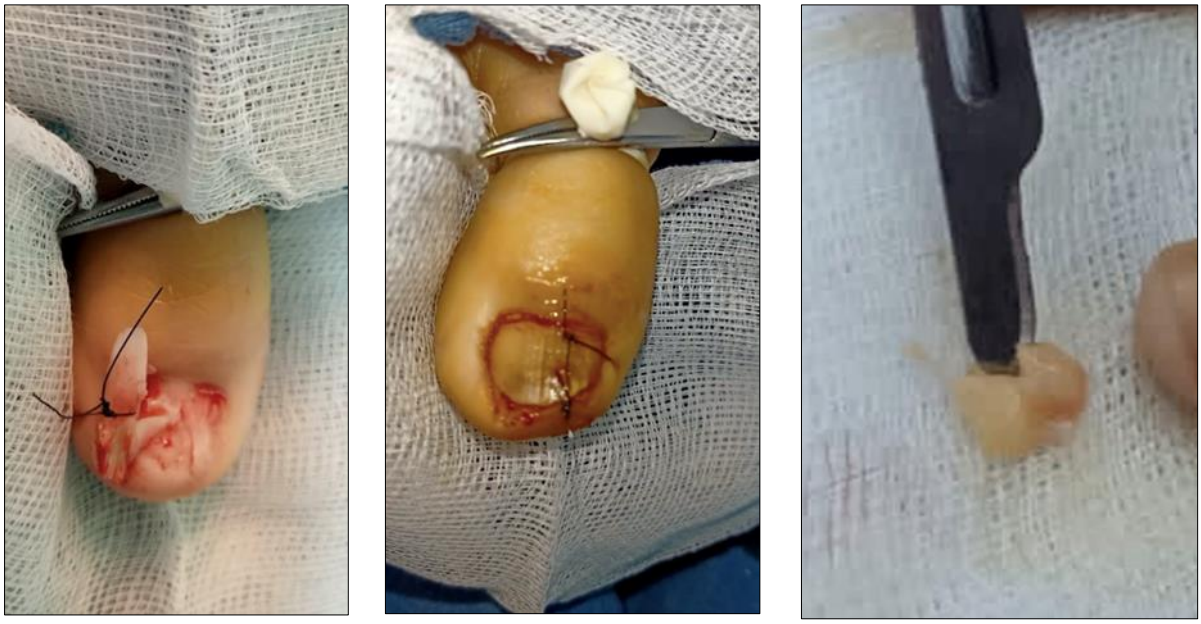

Figure 3 Intraoperative photographs show surgical excision of the tumor and gross specimen following excision

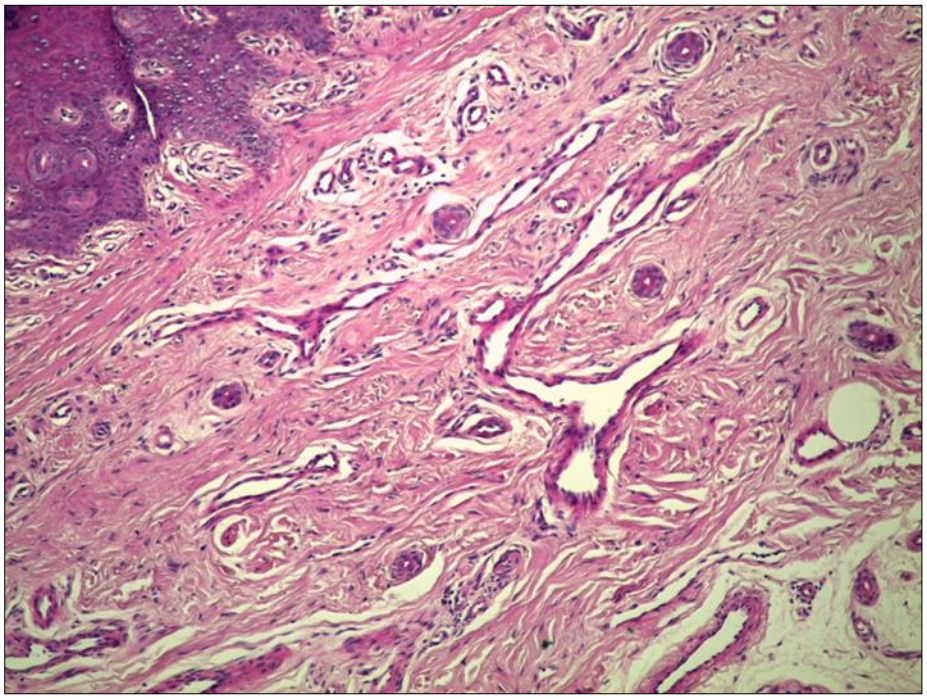

Figure 4 (Coloration HES G * 100) Histologic appearance of the glomus tumor

4. Conclusion

The rarity and unique clinical presentation of glomus tumors pose a diagnostic challenge. Despite the classic triad of severe pain, cold hypersensitivity, and a non-specific examination, modern imaging, particularly MRI, has revolutionized diagnosis by revealing precise tumor location and size. Definitive diagnosis, however, hinges on histological examination. Treatment is solely surgical, requiring a specialized setting and microsurgical techniques. Recurrence is uncommon, and outcomes are generally excellent.

The case report follows scare guidelines [7].

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from the patient.

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