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Review article

Glomus tumours of the hand: Review of literature

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ABSTRACT

Glomus tumours are rare benign vascular neoplasms commonly found in the hand particularly in subungual region. Though, its aetiology remains largely unknown, several hypotheses have been made to explain the etiopathogenesis and cause of pain. These tumours usually present as a bluish or pinkish red discolouration of the nail plate with classical triad of localised tenderness, severe pain, and cold sensitivity. Nevertheless, differential diagnosis of other painful tumours, such as leiomyoma, eccrine spiradenoma, haemangioma, neuroma, osteochondroma, or mucous cyst should always be kept in mind while evaluating a patient with severe pain in the tip of the finger. In addition to the different clinical tests including Love's pin test, Hildreth's test, and trans-illumination test, imaging studies such as magnetic resonance imaging (MRI), ultrasonography, and radiography are often helpful in the diagnosis. Complete surgical excision is a must to get complete relief from the symptoms and to avoid recurrence. Several approaches have been described in the literature. Different surgeons may have different choices and may prefer one approach over the other depending on the anatomical location of the tumours. The purpose of this article is to review the important aspects of glomus tumours in hand concerning their aetiology, clinical presentation, diagnosis, management, and recurrence.

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1. Introduction

Glomus tumours are rare, benign, vascular neoplasms arising from glomus body which is a contractile neuromyoarterial structure found in the reticular dermis. Glomus body consists of afferent arteriole, anastomotic vessel known as Sucquet-Hoyer canal, primary collecting vein, intraglomerular reticulum, and capsular portion.¹ This structure controls blood pressure and temperature by regulating blood flow in the cutaneous vasculature.^{2–4} Hyperplasia in any of these parts can lead to a tumour formation. Although this tumour can be

found anywhere on the body, most common site of its occurrence is distal phalanx of the fingers, especially in the subungual region.⁵ Though this is true in case of female population, males often have these tumours in other parts of the body.⁶

In general, there are two types of glomus tumours, namely, solitary and multiple. Solitary glomus tumours are more common, while multiple glomus tumours rarely occur in the digits.⁶ Multiple glomus tumours have been found simultaneously with type 1 neurofibromatosis and are often painless, making them harder to diagnose correctly.⁶ Magnetic resonance imaging (MRI) is an excellent imaging modality in the

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detection of glomus tumour and also in delineating its anatomical details such as size and location.⁷ Complete surgical excision of the tumour is the recommended treatment to reduce the chance of recurrence.⁸

2. Etiopathogenesis

The aetiology of glomus tumours is unknown and it may be related to sex, age, trauma, or inheritance. Some authors have proposed that a weakness in the structure of a glomus body could lead to reactive hypertrophy after trauma.⁶ Researchers recently reported that a familial variant of glomus tumour had been linked to chromosome 1p21–22 and involved truncating mutations in the glomulin gene, which encoded a 68-kDa protein with unknown function.^{9,10}

Though, the cause of pain in glomus tumour is not clearly understood, several hypotheses have been made; presence of the capsule, which is sensitive to pressure, presence of mast cells releasing substances like heparin, histamine and 5-hydroxytryptamin which render the pressure and thermal receptors sensitive.¹¹ In addition, excessive dominance over the nerve of numerous non-myelinated nerve fibres that penetrate into glomus tumours has been suggested as a cause of pain.¹²

3. Clinical presentation

Glomus tumours account for 1–5% of soft tissue tumours of the hand and 75% of them are subungual in location.¹³ Other less commonly involved sites in the hand are the nail matrix, nail bed, and pulp of a finger.^{1,14} The middle age women are mainly predisposed for these tumours.¹⁵ These tumours usually present as a small, slightly raised, bluish or pinkish red, painful nodule, and when subungual in location, can elevate, deform and discolour the nail (Fig. 1). The typical clinical triad of localised tenderness, severe pain, and cold sensitivity is

Fig. 1 – Bluish pink discolouration of the nail plate of left thumb because of a subungual glomus tumour in a 21 years old female.

highly suggestive of glomus tumour.¹⁶ There is often a history of aggravation of symptoms in cold weather, on holding cold objects in hands, or after placing the hand in cold water.

4. Diagnosis

In addition to the classical presentation, there are three useful tests that help in diagnosing these tumours. In Love's pin test, pressure is applied to the suspected area with a pinhead. The area containing the glomus tumour would be exquisitely painful. Another test is Hildreth's test, in which, a tourniquet is applied along the arm to induce a transient ischaemia. The test would be considered positive if withdrawal of pain from the affected area is noted by the patient. This can be attributed to the temporarily restricted blood supply as it is a vascular tumour. This can be further substantiated by repeating the Love's pin test, which will be painless with inflated tourniquet. On removing the tourniquet, the patient will feel a sudden return of pain. In the third test, which is the cold-sensitivity test, cold water or an ice cube is applied to the affected area.¹⁷ The patients with glomus tumour would feel increased pain in the affected area. Another less commonly used test is the trans-illumination test, in which light is passed through the finger pad.⁶ The tumour will appear as a red opaque image. This test has been found to be 23–38% sensitive and 90% specific.¹⁸

In a study conducted by Netscher et al.,¹⁷ it was found that the Cold sensitivity test had 100% sensitivity, specificity, and accuracy. The Love's pin test was found to be 100% sensitive with 78% accuracy. On the other hand, Hildreth's test was found to be most specific with 100% specificity, 71.4% sensitivity and 78% accuracy.

The differential diagnosis of other painful tumours, such as leiomyoma or eccrine spiradenoma should be kept in mind while evaluating solitary glomus tumours. Moreover, painful tumours such as haemangioma, neuroma or gouty arthritis can simulate the glomus tumour in hand leading to a diagnostic enigma and can pose a therapeutic challenge. Multiple glomus tumours should be carefully differentiated from cavernous hemangioma and blue rubber-bleb nevus syndrome, as they can be easily confused with one another.¹⁹

In spite of the classical presentation, delay in diagnosing these tumours for many years is a significant problem. It can be attributed to the variations in symptom presentation and sometimes obscure symptoms such as chronic pain and hypersensitivity. It is not uncommon that patients are easily misdiagnosed with conditions like neuropathic complaints, arthritis, or neuralgia and may undergo unsuitable treatment including sympathetic ganglionectomy or radicotomy.²⁰ Moreover, its rarity along with lack of suspicion during examination of the patient with impalpable glomus tumour may explain the long delay to reach the correct diagnosis.¹⁶

5. Imaging

Physical examination alone may not be able to diagnose the early lesions of glomus tumour in the hand, especially in the subungual location.²¹ So, the chances of misdiagnosis and

Fig. 2 – Plain radiograph showing bony erosion on distal phalanx of left middle finger because of the paraungual glomus tumour in 25 years old female.

delayed diagnosis are high if relied only on the physical examination.

Pre-operative imaging studies can be useful in case of doubtful diagnosis and also to elicit the anatomical details of the lesion. Radiographs can show cortical thinning or erosive changes in the adjacent bone in some of the cases^{2,14,22} (Fig. 2). Ultrasonography is capable of demonstrating the size, site, and shape of the tumour, but is frequently influenced by the surgeon's experience.^{22,23} Moreover, the curvature of the nail plate may create artefacts in the lateral nail folds, and small and flattened subungual lesions can be difficult to detect with ultrasonography. MR imaging, however, is an excellent imaging modality for detecting the glomus tumour as small as 2 mm.¹⁷ MR imaging can also be helpful in making differential diagnoses, including identifying a number of other lesions such as neuroma, melanoma, pigmented nevus, and haemangioma, as well as foreign bodies.²⁴ The glomus tumour in MRI is characteristically seen as a high signal central dot surrounded by a zone of lower signal intensity¹⁷ (Fig. 3). It shows low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and enhancement on T1-weighted images after gadolinium injection. However, not all glomus tumours will show this nidus appearance.⁵ Some glomus tumours have low signal intensity or isointensity on T2-weighted images and these lesions are better delineated with T2 fat suppression images, with the use of contrast MRI or with magnetic resonance angiography.²⁵ However, the classic MRI findings of a dark lesion on T1 and a bright lesion on T2-weighted images are not specific for glomus tumours. Similar findings can also be seen with cysts and other solid hand tumours^{26,27} although, gadolinium enhanced MRI can help

Fig. 3 – Magnetic resonance images in two different patients (A and B). Glomus tumours showing high signal intensity on T2-weighted images.

differentiate glomus tumours from mucoid cysts and epithelial inclusion cysts because the latter two are nonenhancing. Furthermore, the reliability of MRI scans also needs consideration. In a MR imaging study,⁵ 10% patients out of 40 had negative MRI findings, although histopathology was suggestive of glomus tumours asserting that negative result does not rule out the glomus tumour. In certain cases, ultrasound sometimes scores over an MRI, in certain cases. MRI may mislead these tumours to be cystic lesions unless a contrast imaging is done while an ultrasound is a very good modality to differentiate a solid with a cystic lesion. Moreover, ultrasonography can be a better option than MRI, considering the time required for the test, its cost, and its ability to enable the evaluation of lesions dynamically in real-time.²⁸

Nevertheless, MR imaging can clearly be helpful in diagnosing patients with atypical symptoms and signs. Therefore, the clinical examination should always be combined with the MR imaging for precise and early detection of subungual glomus tumours. This will also help to prevent the misdiagnoses and to excise the tumour in the early stage for earlier pain relief.

6. Management

Complete surgical excision of the tumour is the only effective treatment. Incomplete excision is considered as the main cause of recurrence.^{8,29} Pre-operative MR imaging can be useful to delineate the tumour better and for precise excision reducing the chances of recurrence.¹⁴

7. Surgical approach

In the literature several surgical approaches have been described to excise the tumour. The routinely used traditional approach is direct transungual excision, which consists of cutting through the nail bed to reach the tumour. It affords good exposure to the central subungual tumours, but chances of longitudinal ridge or complete split nail deformity are high in the post-operative period.^{6,30–32}

Multiple modified surgical approaches have been proposed to prevent these complications. Carroll and Berman proposed the use of a “lateral incision close to the edge of the

nail” to approach subungual glomus tumours.¹⁶ It was found to be useful only in case of tumours that are partially under the nail.³³ The Keyser–Littler approach is through a high midlateral incision below the lateral nail fold. In this approach the interosseous structures can get retracted compromising the lateral support to the nail matrix and the nail plate.^{6,33} In a lateral subperiosteal approach, the tumour mass in subungual region is directly accessed without retracting the interosseous structures.³³ However, the degree of exposure is compromised.⁶

In another approach known as modified periungual approach, an L-shaped incision is given over the periungual area, and the tumour is exposed without splitting the nail bed; thus not needing nail bed repair.³² According to some authors, this approach is suitable only for peripheral subungual tumours.^{6,30,32} There can be an inadequate visualisation in case of central subungual tumours with this approach.⁶ In a recently described nail bed margin approach, the nail plate is removed completely to expose the tumour, and a curved incision is made near the side of the tumour along the nail bed margin.³⁴ Dissection is carried down to the distal phalanx, and a dorsal flap containing the nail bed and germinal matrix is raised to expose the tumour sufficiently. According to the authors this approach can expose and completely excise tumours in any subungual region.

In our recently described “Nail-preserving modified lateral subperiosteal approach”, the subungual region is accessed directly without any nail bed dissection or disrupting the interosseous supports of the distal phalanx.³⁵ We use a lateral incision which is curved distally along the pulp of the finger (Fig. 4). On account of the distal curve over the pulp tip, we achieve a large flap to reach to the tumour without any struggle or overzealous retraction may be it in the peripheral or central subungual region. The flap is replaced as it is after the resection of the tumour. The wound healed well in all patients without leaving a noticeable scar mark. Our patients did not have any nail deformity on further follow up.

Histopathological analysis of glomus tumours reveals variable composition of glomus cells, blood vessels, and smooth muscles. Based on this, glomus tumours are categorised into three types: glomangiomas with abundance of vessels; solid glomus tumour chiefly composed of glomus cells; and glomangiomyomas showing a predominance of smooth muscles. Although glomus tumours are essentially

Fig. 4 – Nail-preserving modified lateral subperiosteal approach. A high lateral skin incision is extended distally and curved around the pulp (A) followed by single unit dorsal flap enabling good exposure of the tumour (B).

benign, rarely sarcomas accompany benign glomus tumours to form glomangiosarcoma.¹⁹

8. Factors affecting the recurrence

Almost 4–50% recurrence rate after the surgical excision is noted in the literature. While 'early recurrence' is thought to be a result of incomplete excision¹⁶ or to the presence of a second tumour that was not previously diagnosed and excised during the initial operation³⁶, 'late recurrence' is attributed to the development of a new lesion at or near the excision site.³⁷ The probability of recurrence is higher in case of subungual tumours¹⁷ and this can be attributable to the operative approach and surgeon's tendency to excise the matrix tissue more conservatively to avoid nail plate deformities in the post-operative period.³⁸ Also, large incisions may complicate to large postoperative scars, paresthesias secondary to more injury to small nerve branches, and also to nail dystrophy.^{14,33,38} The skin coloured tumours in contrast to the classical red, blue or purple glomus tumours are difficult to delineate clearly during the surgery.¹⁴ Moreover, their margins are harder to demarcate properly.⁴ These facts are attributable to their incomplete excision, increasing the chance of recurrence.¹⁴ Foucher et al.³⁹ in their study reported 7% recurrence (4 cases) after three to five years. Heim and Hanggi,⁴⁰ in their series, showed that the number of patients with early recurrence was nearly equal to the number of patients with delayed recurrence, with a slight predominance of early recurrences (54%).

Several measures have been recommended to ensure the complete excision and to lower the chances of recurrence. Skin-coloured tumours should be dissected layer by layer, including the entire capsule.³¹ Microscopic monitoring⁴¹ or intraoperative ultrasound may also be useful for ensuring adequate resection.²² A double tourniquet exsanguination procedure (one at midarm level and the other at the base of the digit) has been proposed to better visualise the tumour intra-operatively and according to the authors, none of their patients experienced recurrence of the symptoms during the follow-up period.⁴ In general, if the symptoms of glomus tumours persist for more than 3 months, re-exploration of the affected area and repeat imaging should be done.¹⁷

9. Conclusion

In a nutshell, glomus tumours are the rare tumours with the classical clinical presentation. Differential diagnosis of glomus tumour should always be kept in mind along with other lesions such as haemangioma, neuroma, osteochondroma, or mucous cyst while evaluating a patient with severe pain in the tip of the finger. Complete surgical excision is mandatory to get complete relief in the symptoms and to avoid recurrence. Inadequate excision can be avoided by thorough preoperative assessment, including physical examination and imaging studies and meticulous surgical technique. Several approaches have been described in the literature. Different surgeons may have different choices and may prefer one approach over the other depending on the anatomical location of the tumours. If the symptoms persist or recur after the

surgical excision, high index of suspicion for recurrence of the tumour should be kept in mind and the lesion should be re-explored after repeating the imaging studies.

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Conflicts of interest

The authors have none to declare.

REFERENCES

1. Drape JL, Idy-Peretti I, Goettmann S, et al. Subungual glomus tumours: evaluation with MR imaging. *Radiology*. 1995;195:507–515.
2. Shin DK, Kim MS, Kim SW, Kim SH. A painful glomus tumor on the pulp of the distal phalanx. *J Korean Neurosurg Soc*. 2010;48:185–187.
3. Kale SS, Rao VK, Bentz ML. Glomus tumour of the index finger. *J Craniofac Surg*. 2006;17:801–804.
4. Bhaskaranand K, Navadgi BC. Glomus tumour of the hand. *J Hand Surg Br*. 2002;27:229–231.
5. Al-Qattan MM, Al-Namla A, Al-Thunayan A, Al-Subhi F, El-Shayeb AF. Magnetic resonance imaging in the diagnosis of glomus tumours of the hand. *J Hand Surg Br*. 2005;30:535–540.
6. Samaniego E, Crespo A, Sanz A. Key diagnostic features and treatment of subungual glomus tumor. *Actas Dermosifiliogr*. 2009;100(December):875–882.
7. Takemura N, Fujii N, Tanaka T. Subungual glomus tumor diagnosis based on imaging. *J Dermatol*. 2006;33:389–393.
8. Kim DH. Glomus tumor of the finger tip and MRI appearance. *Iowa Orthop J*. 1999;19:136–138.
9. Boon LM, Brouillard P, Irrthum A, et al. A gene for inherited cutaneous venous anomalies ("glomangiomas") localizes to chromosome 1p21–22. *Am J Hum Genet*. 1999;65:125–133.
10. Brouillard P, Ghassibe M, Penington A, et al. Four common glomulin mutations cause two-thirds of glomuvenous malformations ("familial glomangiomas"): evidence for a founder effect. *J Med Genet*. 2005;42:e13.
11. Lee CH, Byeon JH, Rhie JW, Kang YJ, Cho MJ, Lim P. Clinical analysis of twenty cases of glomus tumor in the digits. *J Korean Soc Plast Reconstr Surg*. 1995;22:169–178.
12. Rodriguez JM, Idoate MA, Pardo-Mindan FJ. The role of mast cells in glomus tumours: report of a case of an intramuscular glomus tumour with a prominent mastocytic component. *Histopathology*. 2003;42:307–308.
13. Tomak Y, Akcay I, Dabak N, Eroglu L. Subungual glomus tumours of the hand: diagnosis and treatment of 14 cases. *Scand J Plast Reconstr Surg Hand Surg*. 2003;37:121–124.
14. Lin YC, Hsiao PF, Wu YH, Sun FJ, Scher RK. Recurrent digital glomus tumor: analysis of 75 cases. *Dermatol Surg*. 2010;36:1396–1400.
15. Moon SE, Won JH, Kwon OS, Kim JA. Subungual glomus tumour clinical manifestations and outcome of surgical treatment. *J Dermatol*. 2004;31:993–997.
16. Carroll RE, Berman AT. Glomus tumours of the hand: review of the literature and report on twenty-eight cases. *J Bone Jt Surg Am*. 1972;54:691–703.

17. Netscher DT, Aburto J, Koepplinger M. Subungual glomus tumor. *J Hand Surg Am.* 2012;37:821–823.
18. Tang CY, Tipoe T, Fung B. Where is the lesion? Glomus tumours of the hand. *Arch Plast Surg.* 2013;40:492–495.
19. Chatterjee JS, Youssef AH, Brown RM, Nishikawa H. Congenital nodular multiple glomangioma: a case report. *J Clin Pathol.* 2005;58:102–103.
20. Lee W, Kwon SB, Cho SH, Eo SR, Kwon C. Glomus tumor of the hand. *Arch Plast Surg.* 2015;42(May (3)):295–301.
21. Koc O, Kivrak AS, Paksoy Y. Subungual glomus tumour: magnetic resonance imaging findings. *Australas Radiol.* 2007;51(Spec No.):B107–B109.
22. Chen SH, Chen YL, Cheng MH, Yeow KM, Chen HC, Wei FC. The use of ultrasonography in preoperative localization of digital glomus tumours. *Plast Reconstr Surg.* 2003;112:115–119.
23. Ham KW, Yun IS, Tark KC. Glomus tumours: symptom variations and magnetic resonance imaging for diagnosis. *Arch Plast Surg.* 2013;40:392–396.
24. Nazerani S, Motamedi MH, Keramati MR. Diagnosis and management of glomus tumours of the hand. *Tech Hand Up Extrem Surg.* 2010;14:8–13.
25. Theumann NH, Goettmann S, Le Viet D, et al. Recurrent glomus tumours of fingertips: MR imaging evaluation. *Radiology.* 2002;223:143–151.
26. Jablon M, Horowitz A, Bernstein DA. Magnetic resonance imaging of a glomus tumour of the finger tip. *J Hand Surg.* 1990;15A:507–509.
27. Peterson JJ, Bancroft LW, Kransdorf MJ. Principles of bone and soft tissue imaging. *Hand Clin.* 2004;20:147–166.
28. Moon ES, Choi MS, Kim MS, et al. Distribution of glomus tumours in fingers. *J Korean Soc Surg Hand.* 2009;14:138–143.
29. Rohrich R, Hochstein L, Millwee R. Subungueal glomus tumours: an algorithmic approach. *Ann Plast Surg.* 1994;33:300–304.
30. Roan TL, Chen CK, Horng SY, et al. Surgical technique innovation for the excision of subungual glomus tumours. *Dermatol Surg.* 2011;37:259–262.
31. Song M, Ko HC, Kwon KS, Kim MB. Surgical treatment of subungual glomus tumor: a unique and simple method. *Dermatol Surg.* 2009;35:786–791.
32. Fong ST, Lam YL, So YC. A modified periungual approach for treatment of subungual glomus tumour. *Hand Surg.* 2007;12:217–221.
33. Vasisht B, Watson HK, Joseph E, Lionelli GT. Digital glomus tumours: a 29-year experience with a lateral subperiosteal approach. *Plast Reconstr Surg.* 2004;114:1486–1489.
34. Wang P-J, Zhang Y, Zhao J-J. Treatment of subungual glomus tumours using the nail bed margin approach. *Dermatol Surg.* 2013;39:1689–1694.
35. Garg B, Machhindra MV, Tiwari V, Shankar V, Kotwal P. Nail-preserving modified lateral subperiosteal approach for subungual glomus tumour: a novel surgical approach. *Musculoskelet Surg.* 2015;100(1):43–48. PMID:25957553.
36. Gandon F, Legaillard P, Brueton R, Le Viet D, Foucher G. Forty-eight glomus tumours of the hand: retrospective study and four-year follow-up. *Ann Chir Main Memb Super.* 1992;11:401–405.
37. Van Geertruyden J, Lorea P, Goldschmidt D, et al. Glomus tumours of the hand: a retrospective study of 51 cases. *J Hand Surg Br.* 1996;21:257–260.
38. Abimelec P, Dumontier C. Basic and advanced nail surgery. (Part 2: indications and complications). In: Scher RK, Daniel III CR, eds. *Nails: Diagnosis, Therapy, Surgery* 3rd ed. Philadelphia: Elsevier/Saunders; 2005:294.
39. Foucher G, Le Viet D, Dailiana Z, Pajardi G. Glomus tumor of the nail area: a series of 55 cases. *Rev Chir Orthop Traumatol.* 1999;85(4):362–366.
40. Heim U, Hanggi W. Subungual glomus tumours: value of the direct dorsal approach. *Ann Chir Main.* 1985;4:51–54.
41. Li QL, Zhang JF, Li HB. Early diagnosis and microsurgical treatment of hand glomus tumor in 30 female cases. *Zhongguo Gu Shang.* 2008;21:381–382 [Chinese].