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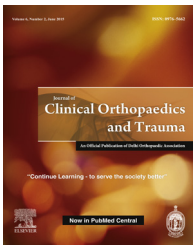

Review article

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.

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 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.

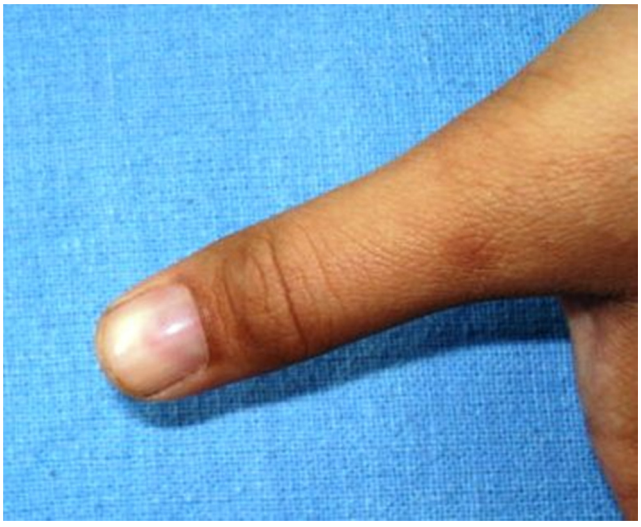

Fig. 1 – Bluish pink discolouration of the nail plate of left thumb because of a subungual glomus tumour in a 21 years old female.

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 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