



# ORIGINAL RESEARCH PAPER

General Surgery

## A RARE CASE OF MALIGNANT GLOMUS TUMOR OF THUMB

KEY WORDS:

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### ABSTRACT

**Introduction:** Glomus tumors are benign perivascular neoplasms of the Glomus body that are involved in the thermoregulation of microvascular blood flow through arteriovenous shunting of blood. These tumors usually occur in areas rich in glomus bodies such as the subungual regions of digits or the deep dermis of the palm, wrist, and forearm. Rarely atypical or frankly malignant Glomus Tumor can occur more frequently as deep-seated, large tumors in the gastrointestinal system, but sparsely in the upper distal extremities. Common symptoms that the patients present with are paroxysmal pain, tenderness and hypersensitivity to cold. Ultrasonography and MRI can be a valuable method of imaging to detect glomus tumors. Radiologically, glomus tumors appear either as bone erosion or invasion depending on where it arises. **Case History** A 83 year old diabetic and hypertensive male patient presented to the Surgery OPD with complaints of ulceration and swelling of his Left Thumb for the past 3 years. He developed an Ulcer in his Left thumb nail which was insidious in onset, gradually progressive and non-healing in nature. The ulcer was associated with pain which was pricking in character, intermittent in nature, non-radiating, and had no aggravating or relieving factors. On examination, an ulcero-proliferative growth was seen on the dorsal aspect of the left thumb, of size 1.5 x 1.5 cm with irregular margins. Edge wedge biopsy of the ulcer was taken, and the HPE showed a poorly differentiated Malignant Glomus Tumour. Upper Limb X-Ray showed no bony involvement. patient underwent partial amputation of the Left thumb under general anaesthesia. The specimen sent for HPE correlated with the clinical diagnosis and showed tumour cells arranged in nests and sheets, as well as congested blood vessels interspersed within the tumour. By IHC the tumour cells were shown to be strongly positive for Vimentin and SOX10. **Conclusion** Malignant transformation of Glomus Tumour is remarkably rare and more commonly reported in lower extremity and abdominal viscera, rather than the upper distal extremities. Glomus tumours tend to pursue a benign course with local aggressiveness; hence it is very essential to pay attention to these cases. We report this case due to its rarity, and emphasize on how vital it is to include it among the numerous differentials for chronic finger pain.

### BACKGROUND

Glomus tumours are rare, usually benign vascular neoplasms that originate from the glomus body and are involved in thermoregulation[1] by altering microvascular blood flow through arteriovenous shunts. They are usually located in the subcutaneous tissue of the extremities. Benign glomus tumours are typically found where glomus bodies are concentrated, such as the digits, palms, wrists, forearms, and feet, especially in the subungual layer of the fingers. Malignant glomus tumour also known as glomangiosarcoma on the other hand are exceedingly rare, and only 6 reports of malignant glomus tumour in the hand has been reported [2]. According to a reclassification of glomus tumors, a diagnosis of malignant should only be given for tumors that have the potential to spread; these characteristics appear to be correlated with a deeper location, a size larger than 2 cm, atypical mitotic figures equal to or greater than 5 mitotic figures per 50 high-power field, and a nuclear grade that ranges from moderate to high[3]. We present to you a case of a malignant glomus tumour in the thumb of a patient, where such a tumour seldom presents.

### Case Presentation

An 83 year had come with complaints of pain in his left thumb followed by swelling which ulcerated over past 3 years. The swelling and ulcer were insidious in onset, gradually progressive and the ulcer was non-healing in nature. The pain was intermittent, non-radiating, aggravated on cold exposure and of pricking type. On examination, a swelling with ulceration was seen on the dorsal aspect of the left thumb, of size 1.5 x 1.5 cm with irregular margins (figure 1), no active bleeding was noted and on palpation severe tenderness was elicited and no palpable axillary or epitrochlear lymph nodes.

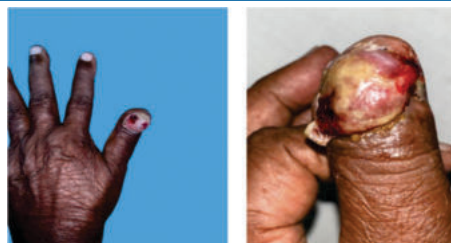


Figure 1

### Investigation

USG Left Axilla was done and showed no lymphadenopathy, no significant findings were noted. X-Ray Left Hand (figure 2) showed no bony involvement. Edge wedge biopsy of the ulcer was done, and the HPE and IHC showed features of a poorly differentiated malignant glomus tumour (figure 3). Other routine basic investigation were unremarkable.



Figure 2

### Treatment

Patient underwent amputation of the left thumb (figure 4), under general anaesthesia. The specimen was sent for Histopathological examination.

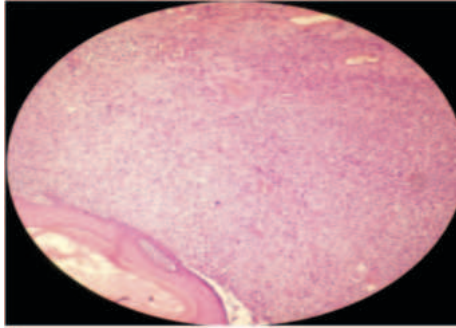


Figure 3



Figure 4

### Outcome And Follow Up

Histopathological examination revealed tumour cells arranged in nests and sheets with congested blood vessels interspersed within the tumour and atypical mitotic figures were found to be 5/50 HPF with vesicular nuclear chromatin and moderate cytoplasm. (Figure 5) .The margins were negative for malignancy.

### Immunohistochemistry:

IHC was strongly positive for Vimentin and SOX10, consistent with the diagnosis of malignant glomus tumour. At follow up after 6 months wound was found to be healthy and no recurrence elsewhere noted.

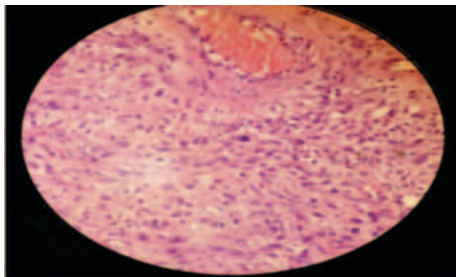


Figure 5

### DISCUSSION

The glomus tumor is a rare benign neoplasm that arises from the neuroarterial structure called glomus body [4], which accounts for 1 % to 4.5 % of tumors in the hand. The normal glomus body is located in the stratum reticular throughout the body, but is more concentrated in the digits[5] . Their main function is thermal regulation. The average age of presentation is from 30 to 50 years of age, although can occur at any age.Average time from onset of symptoms to the correct diagnosis is seven years.Glomus tumours remain notoriously difficult to diagnose, with extreme pain suffered by patients for long periods of time before symptom resolution, as in this patient's case. Carroll and Berman described a classical triad of symptoms for benign Glomus tumours that they felt were pathognomonic: 'spontaneous lancinating pain, extreme pain with the gentlest of touch, and intolerance to changes in temperature'. Clinical tests such as

the Love test (point tenderness) [6] and Hildreth's sign (decreased pain on exsanguination of the limb and applying a tourniquet) have shown to be highly suggestive of glomus tumours. In lesions affecting the distal phalanx, plain radiography most frequently reveals osteolytic lesion with sclerotic border or bone erosion.Glomus tumors as small as 2 mm can be highly sensitively detected by color-duplex ultrasonography[7,8,9].When the precise location of the lesion or the diagnosis is unknown, MRI may be helpful [10,11].Subungual lesions appear as a black, well-defined mass on T1 MRI imaging, although a high homogenous signal is seen over the tumor on T2-weighted images [12]. Sequences measuring fat saturation and postgadolinium may help distinguish the lesion even further.This pattern of magnetic resonance signal can be observed in any type of vascular tumor, including minor subungual or digital lesions; nonetheless, it is thought to be pathognomonic for glomus tumors.The usual course of treatment for glomus tumors is still surgical removal, with well-defined margins signifying the effectiveness of the procedure. The glomus tumor's hallmark histological characteristics include homogeneous, angiocentric sheets of cells with oval nuclei that surround arteries to form a perivascular "collar."Malignant glomus tumors, also known as glomangiosarcomas, are rare soft tissue tumors[13] .They have been reported in many different anatomic locations with the lower extremity and abdominal viscera being the most common. Their location in the hand is extremely rare[14] . Glomangiosarcomas tend to pursue a benign course with local aggressiveness . However, if malignant features are suggested on histology as in this case, a wider excision is required as well as close follow-up review to assess for metastases. Some of the subungual lesions that need to be kept in mind during evaluation of these patients include benign solid tumors (glomus tumor, subungual exostosis, soft-tissue chondroma)[15] benign cystic lesions (epidermal and mucoid cysts), and malignant tumors (squamous cell carcinoma and malignant melanoma).

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