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Original Research Paper

Plastic Surgery

GLOMUS TUMOR IN THE PULP OF FINGER – A RARE PRESENTATION AND DIAGNOSTIC PEARLS

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ABSTRACT **Summary:** Glomus tumors are rare and occur typically in the subungual region of the digits of extremities. They present with severe pain out of proportion to the size of the tumor and are associated with severe cold intolerance. Possibility of glomus tumour must be considered in the differential diagnosis for evaluation of unexplained nocturnal or rest hand pain. We present a unique case of 73-year-old male patient treated for rare presentation of the tumor at the pulp of finger. We also discuss clinical, histopathological and immunohistochemistry diagnosis and management of glomus tumors of Hand.

KEYWORDS : Glomus Tumour, Glomangioma, Glomangiomatosis, Perivascular Neoplasm, Sub-ungual Glomangiomyoma, Sucquet-hoyer Canal.

BACKGROUND:

Glomus tumours are benign perivascular neoplasms that originate from glomus bodies and comprise just 1% of tumours arising in the hand, with fewer than 10% involve the volar pulp of digits [1,2,3]. In the hand they are often found in the subungual regions, as well as in the nail matrix, nail bed and involvement of pulp of a finger is very rare. An exophytic glomus tumour on the pulp of finger in pregnant women has been reported in literature. The period of evolution of glomus tumour ranges from 1 to 7 years. Glomus body is an innervated coiled arteriovenous dermal shunt that normally control blood pressure and skin temperature through arterio-venous shunting of blood. Glomus tumour arises from the arterial end of the glomus body. They are relatively uncommon and usually less than one centimetre in size. Patients typically present with a triad of symptoms including excruciating paroxysmal pain that is worse at night, out of proportion to tumour size, cold sensitivity and severe point tenderness[4].

Glomus tumours are benign lesions arising from modified smooth muscle cells of neuromyo-arterial component of glomus body. Histologically they resemble glomus bodies. They are found more in subungual regions of digits, in the subcutis and superficial soft tissues deep dermis of the palm, wrist, and forearm [5]. Glomus tumour appear as small (less than 1cm) blue-red nodule and may occur in different viscera including the lung, liver stomach, pancreas, gastrointestinal, and genitourinary tract[6]. Subungual presentation has a female preponderance and there is no sex predilection evident for presentation in other locations. Glomus tumours are typically composed of 3 components: glomus cells, smooth muscle cells, and vasculature. The classical histological features of the glomus tumour include angiocentric uniform sheets of cells with oval nuclei, forming a perivascular "collar" around vessels[7]. Atypical or frankly malignant glomus tumours are exceedingly rare and more frequently are deep-seated large tumours in the gastro-intestinal system. Glomangioma and glomangio-myoma are classic variants of the common form of glomus tumours.

Case presentation:

A 73 years old right handed male working as a security guard presented to Hand surgery outpatient Department with history of pain in the pulp of his right mid finger of 6 years duration. There is no history of trauma. He is not diabetic, not hypertensive and does not have any other medical condition. Initially the patient noticed cold intolerance and then developed lancinating type of pain in the digit. On clinical examination of right middle finger there is no visible swelling. Palpation elicited severe tenderness on the pulp of the terminal phalanx, slightly ulnar to midline and on deep palpation a tender, diffuse, palpable mass measuring 0.5 centimeter was identified.

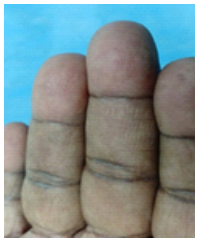

Figure 1: There was no obvious swelling in the pulp of middle finger.

Clinical diagnosis and differential diagnosis:

A clinical diagnosis of glomus tumour was considered along with other differential diagnosis including the possibility of vascular or melanocytic lesions and lipoma.

Investigations:

Laboratory diagnosis for screening tests were all negative. Plain X-ray of the right hand was taken.

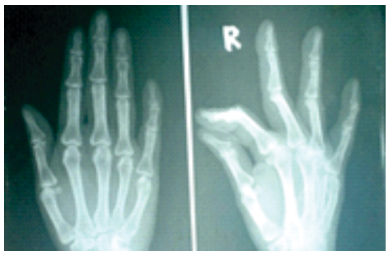

Figure 2: Plain radiography findings were unremarkable.

Treatment: With a clinical diagnosis of glomus tumour the patient was taken up for excision biopsy of the lesion. The surgery was done under loupe magnification, under digital block anaesthesia and tourniquet control using a tourniquet. A curvilinear incision with convexity to ulnar side of terminal phalanx of right middle finger was made to avoid scar on the contact radial border of finger.

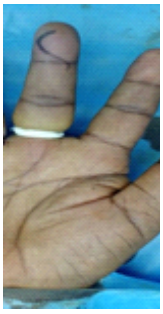

Figure 3: A finger tourniquet was used at the base of the middle finger.

The tumour was found to be deep in the pulp of terminal phalanx of mid finger slightly to the ulnar side of midline.Tumour was excised in toto and skin wound was closed primarily using 4-0 ethilon sutures. Post-operative period was uneventful. The wound healed well and sutures were removed on seventh post-operative day. He was adviced to follow scar care treatment protocol.

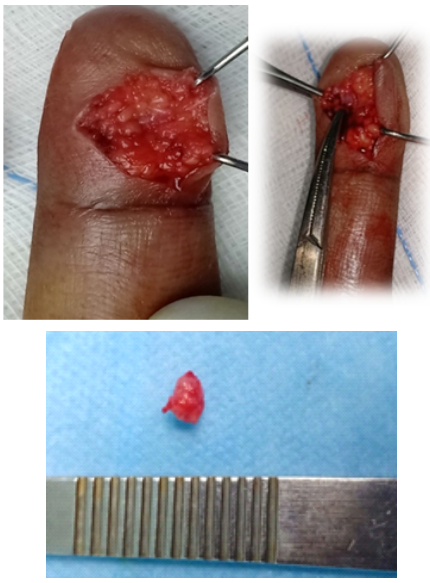

Figure 4: The tumour was deep in the pulp just above terminal phalanx. The excised specimen measured 1 x 0.5 cm.

Pathological diagnosis:
Histopathological study confirmed diagnosis of glomus tumour. Multiple fragments of fibro-adipose and fibro-collagenous tissue shows well circumscribed tumour composed of predominantly glomus cells, blood vessels and smooth muscle cells. The glomus cells are arranged in sheets interspersed with myxoid stroma showing round sharply punched out nucleus with indistinct border, eosinophilic cytoplasm and homogenous nuclear chromatin and few of them showing prominent nucleoli. No nuclear atypia or mitosis seen. It was recommended immunohistochemistry confirmation with vimentin, SMA and Ki67. IHC was positive for SMA and Vimentin and the Ki67 labelling Index was 1%. Based on H & E and IHC studies, diagnosis of glomus tumour was confirmed.

Analysis: The following validated outcome questionnaires were completed prior to treatment and prospectively post-treatment.

Numeric Pain Rating Scale: The patient rated his pain over the previous week on a scale of 0–10. **Quick disability of arm, shoulder and hand** (QuickDASH): The DASH outcome measure is a validated quality of life measure used to assess disability secondary to upper limb complaints. In this case study, the QuickDASH is used for assessment consisting of a disability Score. The score ranges from 0 (no disability) to 100 (most severe disability). An improvement of 10 points is considered a minimal clinically important difference.

OUTCOME AND FOLLOW-UP: The patient was followed in the out-patient department. The patient experienced significant relief from symptoms of local and paroxysmal pain. The scar settled well without any pain sensation. He has returned to his job and has not experienced intolerance to cold.

DISCUSSION:

Glomus tumours, also known as Barré–Masson syndrome, are rare benign hamartomas that arise from the normal glomus apparatus, located in subcutaneous tissue. Wood was the first to describe the condition in 1812 as a painful subcutaneous tubercle. Masson in 1924 coined the descriptive term "glomus" (Latin for "ball") and with histopatho-logical study he recognised their neuromyoarterial origin. Touraine in 1936 first described multiple hereditary glomangiomas. Glomus body consists of an afferent arteriole, an anastomotic Sucquet–Hoyer canal, an efferent venule, the intraglomerular reticulum and its capsule. The Sucquet–Hoyer canals are lined by endothelial cells which are surrounded by smooth muscle cells and large cuboid glomus cells are interspersed in the smooth muscle[8]. With application of tourniquet to the finger, glomus tumours are characterized by a capsule surrounded by dark red, beige-pink colour. They are either solitary or multiple. Solitary glomus tumours are far more common than the multiple variant. According to Rettig and Strickland, the solitary variant is mostly found in the hand, with 25%–75% occurring in the subungual region and are more frequent in women in the age group of 25- 40[9]. Multiple glomus tumours are most often are painless, pink or purple appearance with a nodal shape [10,11]. They develop in young children or in males in an autosomal dominant fashion. Glomus tumour is also classified as digital and extra-digital.

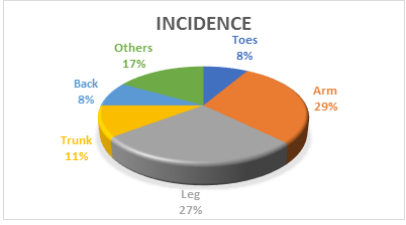

Figure 5: Incidence of glomus tumour in different body parts.

Clinical tests:
(1) Love test: Apply pressure on the tumour with a blunt object to localize the tumour. Positive Love's pin test means patient experience severe pain when skin overlying the tumour is pressed with ballpoint pen or a pinhead. (2) Hildreth's test: Elevate patients' arm, exsanguinate it and a tourniquet is inflated to 250 mm Hg. Pain and tenderness are reduced on palpation of the tumour. Positive test is sudden onset of severe pain and tender-ness in the area of tumour when the cuff is released[12]. (3)Provocation test shows cold impact on pain. The cold-sensitivity test is positive when hand immersion in cold water elicits severe pain in and around the lesion. These clinical diagnostic tests are helpful, but still have limitations in accurate diagnosis. Love's test has a sensitivity of 100% and 78% accuracy. Hildreth's test has a sensitivity of 71.4%, a specificity of 100% and accuracy of 78% and the cold-sensitivity test has a sensitivity, specificity and accuracy each of 100%. (4) Recently, EKIN et al introduced a Transillumination test useful in the diagnosis and location of sub-ungual tumours.

Differential diagnosis

include vascular or melanocytic lesions and Lipoma. There may be confusion and difficulty in differentiating cyst, angioma, fibroma, neuroma, melanoma,nevus, leiomyoma, pyogenic granuloma and spiradenoma.

Mechanism of pain in Glomus tumour:

Hypertrophy of glomus body is the feature of glomus tumour. Glomus cells are specialized perivascular muscle cells that are round or oval and have a dense, granular cytoplasm. The causes of pain are due to

- The capsule of the tumours are more sensitive to pressure. Temperature changes cause contraction of myofilaments as a response with increase in intracapsular pressure with resultant severe pain.
- Abundant mast cells in glomus tumours release substances such as heparin, 5-hydroxytryptamin, and histamine. This results in receptors to pressure or cold stimulation to be sensitive[13] and
- Lancinating type of pain is due to non-myelinated nerve fibres, intermixed with thick-walled capillaries. Numerous non-myelinated nerve fibers penetrate into glomus tumours.

Typical histological features: Glomus tumours are typically composed of 3 components: glomus cells, smooth muscle cells and vasculature. The classical histological features of the glomus tumour include angiocentric uniform sheets of cells with oval nuclei, forming a peri-vascular "collar" around vessels. Three different tumour variants are differentiated by their histological characteristics[14,15].

- Solid form(more common)
- Glomangioma and glomangiomyoma(classic variants)
- Malignant form: Glomangiosarcoma

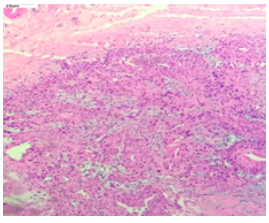

Figure 6: Section showing a well circumscribed lesion with thin walled blood vessels surrounded by uniform cells (100 X H&E).

Glomus cells have both smooth cell and pericyte phenotype as confirmed by immunohisto-chemical and electron microscopic analyses. Glomus tumours are characteristically and diffusely immune-reactive for Smooth Muscle Actin (SMA), Muscle Specific Actin(MSA), and h-Caldesmon. Vimentin and collagen type IV are also expressed, although they are non-specific. Variable expression of CD34, and to a lesser extent desmin, has also been reported. ***Pathology of glomus tumour is confirmed from hematoxylin and eosin stains and are positive for actin, CD31, CD34, SMA, ETS-related gene and vimentin***[16]. They are negative for S100 and Desmin usu. The following guidelines are helpful in immunohistochemical and electron microscopy studies.

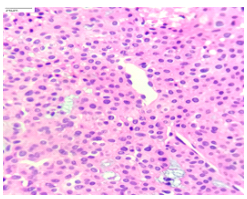

Figure7: High Power showing glomus cells with uniform round to oval cells, moderate amount of eosinophilic cytoplasm and homogenous chromatin 200 X H&E

Positive stains: Vimentin (100%), smooth muscle actin (99%), muscle specific actin (95%), calponin (80%), CD34 (32 - 53%; typically focal); Collagen type IV, laminin (91%; peri- cellular), h-caldesmon (87%).

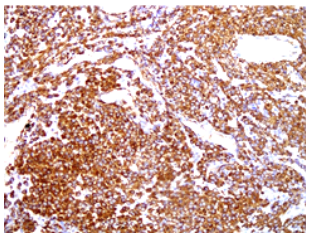

Figure 8: IHC for Smooth muscle Actin (SMA) showing strong postivity in the glomus cells X100.

Negative stains include cytokeratin, CD31, S100, HMB45; CD117, desmin, chromogranin, synaptophysin, CD20, CD45 and WT1[17,18]. **Electron microscopy** shows thick basal lamina surrounds individual glomus cells except at cellular junctions. Pinocytic vesicles and myofibrils with dense bodies in the cytoplasm are characteristic features[19].

CONCLUSION:

Glomus tumour is a rare perivascular benign tumour arising from the Sucquet-Hoyer canal of the normal glomus body, most commonly in the digits. We report a rare case of glomus tumour at pulp of the finger, with long-term severe pain, cold sensitivity, and point tenderness. Glomus tumours are diagnostic challenge as they are small and situated deep in the fingertip. Awareness regarding the unusual locations of tumours such as the pulp of the fingers is important for early treatment. Patients presenting with a typical triad of symptoms should be evaluated for glomus tumour. Ultrasonography or MRI scan should be performed to ensure proper diagnosis and treatment [20,21]. Histopathology revealed a well-circumscribed tumour composed of clusters of monotonous polygonal cells surrounding capillary-sized blood vessels. Tumour cells also showed immunopositivity for smooth muscle antigen and vimentin. Finally, complete surgical excision must be meticulously planned for permanent cure and recurrence rate can be anywhere from 5% to 50% due to incomplete excision.

Learning points:

- A high index of suspicion of glomus tumour is essential for clinical diagnosis.
- Glomus tumour should not be confused with hemangioma or para-ganglioma. Patients with unexplained paroxysmal shooting pain nocturnal or rest hand pain, sensitivity to cold, and localized tenderness in the finger must be investigated to rule out glomus tumour.
- Various presentations of glomus tumours and occurrence at rare locations must always be kept in mind.
- Imaging studies like ultrasonography and magnetic resonance imaging are needed for confirmation.
- Incision should be planned to avoid scars on the contact areas of the fingers.
- Immunohistochemistry(IHC) studies are essential to confirm diagnosis and also to rule out the possibility of malignant transformation, which is extremely rare.
- The standard treatment of choice is surgical removal. Recurrence of glomus tumour generally resulted from incomplete excision.

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