Glomus tumor of hand: A rarity treated by transungual approach

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Abstract

Introduction: Glomus tumor is a benign and rare vascular tumor arising from the glomus body, commonly found in the hand particularly in subungal region. These tumors present usually as bluish discoloration of nail plate with classical triad of excruciating pain, localized tenderness and sensitivity to cold. It’s etiology remains unknown but several hypothesis explain the excruciating pain. Different clinical test including Love’s pin test, Hildreth’s test and trans-illumination test are often helpful in diagnosis. MRI (Magnetic Resonance Imaging) is an excellent imaging modality in the diagnosis of these lesions. Diagnosis of tumor is delayed because of subtle clinical signs and also surgeons often miss advising MRI to patients due to its high cost. There are two main approaches for excision of glomus tumor-transungual and perungual. Selection of approach depends upon surgeon’s choice and anatomical location of the tumor. Complete surgical excision of the tumor reduces chance of recurrence. The aim of this study is to review the time from onset of symptoms to the diagnosis, management of problem and recurrence retrospectively.

Material and methods: This study was done retrospectively between May 2017 to May 2019 at tertiary care center of eastern India. All patients with histopathologic ally confirmed subungal glomus tumor of distal phalanx of hand were included in this study. Patients with Reynolds disease were excluded from this study. MRI was done preoperatively and histopathological analysis of the excised specimen was done postoperatively to confirm the diagnosis. Patients were followed-up after every 3 month for 1 year to see any complication and recurrence.

Results: It was found that the patients had clinical symptoms for an average of 2.3 years. All patients were women with mean age of 34.2 years. Out of 5 patients, 2 patients developed nail deformity. Recurrence was not found till last follow-up period. All patients were returned to their normal routine work and regained their full function of the hand.

Keywords: Glomus tumor, Hildreth’s sign, love’s sign, subungal, transungual

Introduction

Glomus tumor is a rare benign hamartoma of vascular origin arising from the glomus body [1]. It represents 1 to 5% of all tumors of hand [2]. Masson in 1924 was the first scientist to give its histological details describing it as neuro-myoarterial tumor [3]. Histologically, it is composed of three main structures namely glomus cell, the vasculature and smooth muscle cell. Hyperplasia of any of these structures result in a glomus tumor [4]. Most of the time it is located in the subungal region [5] but can occur in any part of our body. The pulp and the parungual regions are other common places. It classically affects women between 20 to 40 years [6]. Normally, its main function is to regulate the normal skin temperature [7]. Generally, patient presents with excruciating pin point pain under the nail which doesn’t get relieved on taking analgesics. This often causes misdiagnosis such as neuropathies, neuralgia, arthritis, chronic paronychia, subungal exostosis, raynaud’s phenomenon and patients keep on visiting different hospitals in pursuit of relief. The pathophysiology is attributed to the presence of mast cells which release substances like heparin, 5-hydroxytryptamine and histamine. These substances stimulate pressure and cold receptors [8]. The presence of innervation is responsible for pain [9]. MRI is an excellent imaging modality in the diagnosis and locating the tumor but many surgeons often miss advising MRI to their patients because of subtle clinical signs and also because of its high cost. MRI is still beyond the reach of economically weaker part of the society. Surgical excision either by transungual or perungual route is the only known treatment option [10].
Although glomus tumor is a benign tumor but sarcomatous changes has been seen as an exception[11].

Material and Methods
It was a retrospective study carried out in a tertiary care hospital in eastern India between May 2017 to May 2019. Data was obtained retrospectively from hospital records and the patients were called for clinical follow-up. 5 patients with histopathologically confirmed subungual glomus tumor of distal phalanx of hand were included in this study. Patients presenting with Raynaud’s syndrome were excluded. All patients were female. The mean age of patients was 34.2 years (range 28-42 years). The right hand was affected in 3 patients and left hand was affected in 2 patients. The index finger was the most common digit to be involved (n=3) followed by ring and middle finger (n=1 each) [Table 1]. The mean duration of symptoms before diagnosis was 2.3 years (range 1-3 years). All patients had pain in the nail bed and 3 patients had sensitivity to cold. Clinically, tenderness was assessed by the Love’s pin test [12] (the patient experiences severe excruciating pain when the nailplate over the lesion is pressed with a pin head or tip of a ball point pen) which was positive in all patients. The Hildreth’s test[13](disappearance of pain and tenderness over the lesion after application of a tourniquet cuff proximally in the arm) was also positive in all patients. MRI scan was done preoperatively to identify the lesion [Fig.1]. Informed consent was taken from all patients before surgery. Surgical excision of tumor was done under local anaesthesia (2% xylocaine) using ring block technique. Transungual approach was used in all patients. The nail plate was carefully elevated without damaging the geriminal matrix of nail. The tumor was identified by bluish –red discoloration in the nail bed. A vertical incision was made in the nail bed over the tumor. The nail bed was retracted and tumor was excised completely [Fig. 2]. Haemostasis was achieved. The nail plate was repositioned without suturing over the nail bed and compression dressing was done over the finger. Histopathological analysis was done postoperatively to confirm the diagnosis. Check dressing was done on 3rd day. The nail plate usually fell off in 4-6 weeks and new nail was completely formed in 8-10 weeks. Patients were followed-up after every 3 month for 1 year to see any complications and recurrence.

Table 1: Clinical details of the patients

| Age(years) | Sex | Side | Finger | Symptoms                  | Duration | Signs                     | Histopathology | complications |
|------------|-----|------|--------|---------------------------|----------|---------------------------|----------------|--------------|
| 28         | F   | Rt   | Index  | Pain, cold sensitivity    | 1 year   | Lv(+ve), Hd (+ve), Localised tenderness (+ve) | Nil            |              |
| 35         | F   | Lt   | Index  | pain                      | 3 years  | Lv(+ve), Hd(+ve), Localised tenderness (+ve) | Nail deformity |              |
| 30         | F   | Lt   | Index  | Pain                      | 2 years  | Lv(+ve), Hd(+ve), Localised tenderness (+ve) | Nil            |              |
| 36         | F   | Rt   | Middle | Pain, cold sensitivity    | 2.5 years| Lv(+ve), Hd(+ve), Localised tenderness (+ve) | Nail deformity |              |
| 42         | F   | Rt   | Ring   | Pain, cold sensitivity    | 3 years  | Lv(+ve), Hd(+ve), Localised tenderness (+ve) | Nil            |              |

Discussion
Glomus tumor arises from glomus body. It is rare, benign, and vascular neoplasm. The glomus body is an arterio-venous anastomosis found beneath the nails and on ventral aspect of tip of finger. This glomus body controls blood pressure and temperature by regulating blood flow in the cutaneous
vascular [7, 12]. The middle age women are mainly predisposed for these tumor [14]. Grossly, these tumors present as a small, slightly raised, bluish or pinkish red, painful nodule, and when subungual in location can elevate, deform and discolor the nail. Microscopically, the lumen of the anastomotic vessel is tortuous and dilated, lined by a single endothelium layer. This layer is supported by fibrous layer, which is surrounded by cuboidal or rounded endothelial cells - the glomus cell. A collagen fibre stroma is present between the endothelial cells, which contain nerve fibres [15]. Infiltration of surrounding tissue is not seen. An incomplete capsule surrounding the lesion is present [16]. This tumor account for 1-5% of soft tissue tumors of hand and 75% of them are subungual in location [2-5]. In our series, all the 5 tumors were subungually located. The tumor is most commonly seen between 20-40 years of age, with women being affected 4 times more than men [6, 14]. The mean age in our series was 34.2 years and all were women. Various studies show that a long duration of symptoms with various erroneous diagnosis seems to be the rule rather than exception [3, 12, 16, 17]. In our study, the mean duration of symptoms was 2.3 years (range 1-3 years). The two largest reported series of subungual glomus tumor emphasize the importance of clinical diagnosis based on the clinical triad of excruciating pain, localized tenderness, and sensitivity to cold [3, 16]. In our series, all patients had pain and localized tenderness but only 3 had sensitivity to cold. The Love’s test and Hildreth’s test were positive which are considered “virtually pathognomonic” of glomus tumor [12, 13]. Bluish or pinkish red discoloration was visible through nail plate in 26-50% of the subungal lesions [3, 5, 16]. In our series 3 patients had visible discoloration under nail. In subungual lesion, nail deformity can occur [3]. Patients kept on visiting one hospital to another in pursuit of relief before coming to us with incomplete investigation and treatment because this condition can be misdiagnosed as other painful tumors like leiomyma, haemangiona, neuroma or gouty arthritis, chronic paronychia, subungual exostosis and raynaud’s phenomenon [18]. To confirm the diagnosis of subungual glomus tumor, we rely only on clinical tests and MRI. MRI helps to locate the tumor in nail bed. It appears slightly hypointense on T1 and hyperintense on T2 [19]. Surgical excision is the treatment of choice for subungual glomus tumor [3, 14, 16]. It was done through transungual approach under low anaesthesia (2% xylocaine) using ring block technique in all the cases which is recommended by Carroll and Berman and Van Geertruyden et al. [3, 16]. The nail falls off over a few weeks and takes 8 to 10 weeks for appearance of new nail. Periungual approach, Keyser-Littler approach and Nail-preserving modified lateral subperiosteal approach is the other known treatment option [18]. Surgical complications are nail deformity and recurrence of tumor. Chances of longitudinal ridge or complete split nail deformity are high with the transungual approach [15, 18]. The incidence of nail deformity after this approach varies from 3.3% to as high as 26.3% [3, 20]. Almost 4-50% recurrence rate after the surgical excision is noted in the literature. While ‘early recurrence’ is thought to be due to of incomplete excision [16] and ‘late recurrence’ is attributed to the development of a new lesion at or near the excision site [3]. True recurrence of the tumor is rare [16]. We had not found any recurrence during follow up period of 1 year. The limitation of our study is that since it is rare tumor, very few cases formed a part of this study.

Conclusion
Glomus tumor is a rare entity and they frequently escape diagnosis because these are small in size and have subtle clinical signs. So, a high index of suspicion and awareness is needed for a proper and timely diagnosis. History and clinical examination forms the core part of diagnosis. MRI should be routinely performed for proper location of the tumor. Surgical excision is the only treatment option available.

Abbreviation: F = Female, Rt = Right, Lt = Left, Lv = Love’s test, Hd = Hildreth’s test, (+ve) = positive

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