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***Case Report***

# A RARE CASE OF EXTRA DIGITAL GLOMANGIOMA

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## ABSTRACT

Glomus tumors are relatively rare vascular tumors with reported incidence of 1.6% of all soft tissue tumors. It is variously regarded as a hamartoma or a neoplasm of neuromyoarterial glomus, which consists of dilated vascular channels, surrounded by proliferating glomus, nerve cells, and plays an important role in temperature regulation. We report the case of glomangioma in a 31-year-old male who came to surgical OPD with a painful single swelling in the right middle third of forearm of nineteen years duration. on examination a single round swelling of about 3x2 cms in size on the flexor aspect of the right middle third of forearm. The swelling was soft in consistency and tender with sign of compressiblity. the ultrasound revealed well defined highly vascular soft tissue lesion in the subcutaneous plane of the right posterior forearm arterio venous malformation to be considered. The excision biopsy was done and histo pathological examination revealed well circumscribed lesion composed of ecstatic and irregular vascular channels lined by round glomus cells with distinct intercellular junctions. The above features were consistent with glomangioma. It is being presented for its rarity.

***Keywords:*** *Glomangioma, Glomus Tumor, Soft Tissue Tumor, Glomus Cells*

## INTRODUCTION

Glomus tumours are relatively rare vascular tumours with reported incidence of 1.6% of all soft tissue tumours (Shugart *et al.,* 1963). It is variously regarded as a hamartoma or a neoplasm of neuromyoarterial glomus, which consists of dilated vascular channels, surrounded by proliferating glomus and nerve cells (Suckquet-Hoyer canal) and plays an important role in temperature regulation.

Solitary glomus tumours are usually seen in adults equally commonly in both sexes except for subungual glomus tumours which show a female preponderance (Shivaswamy *et al.,* 2003). A soliltary glomus tumour is a pink or purple nodule with classic triad of pain, cold sensitivity and point tenderness (Walsh and Eady, 2004). The commonest site is the hands, particularly the fingers. Here we report a case of Glomus tumour involving unusual site.

## CASES

A 31-year-old male came to Surgical OPD with a painful swelling in the right middle third of forearm of nineteen years duration. The swelling started as a small one gradually increased over a period of nineteen years to attain the present size of 3x2 cms associated with pain on and off especially while giving pressure. There was no history of trauma preceding the swelling. On examination a single round publish blue swelling of about 3x2 cms sized on the flexor aspect of the right middle one third of the forearm (Figure 1.1). The swelling was soft in consistency and tender.

While giving compression it was completely disappeared. On ultra sonagram of the right forearm reavealed a well defined highly vascular soft tissue lesion in the subcutaneous plane of the right posterior forearm-AVM to be considered (Figure1.2).

All the basic blood investigations were normal. The excision biopsy was performed under local Anaesthesia and specimen sent for histopathological examination (Figure 1.3) Macroscopically a single congested nodular tissue of measuring 1.5x1.3x0.8 cms. On cut section it was grey brown spongy. Microscopically a well circumscribed lesion composed of ectatic and irregular vascular channels lined by round glomus cells with distinct intercelluar junctions. No evidence of malignancy (Figure1.4). The above features were consistent with the diagnosis of glomangioma.

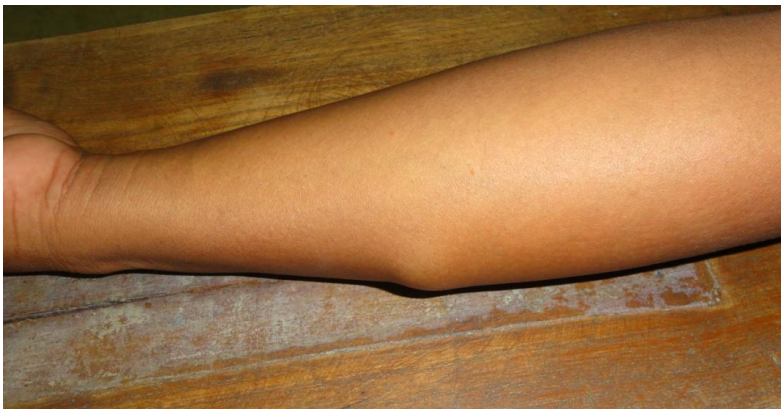

**Figure 1.1: Shows a single swelling in Rt middle third of forearm**

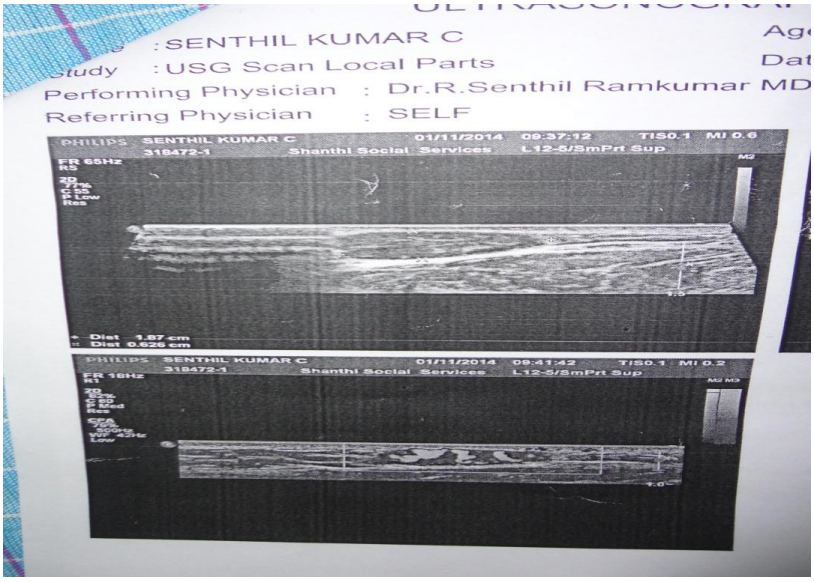


**Figure 1.2: Shows the image of USG with a well defined highly vascular soft tissue lesion in the subcutaneous plane of the right posterior forearm**

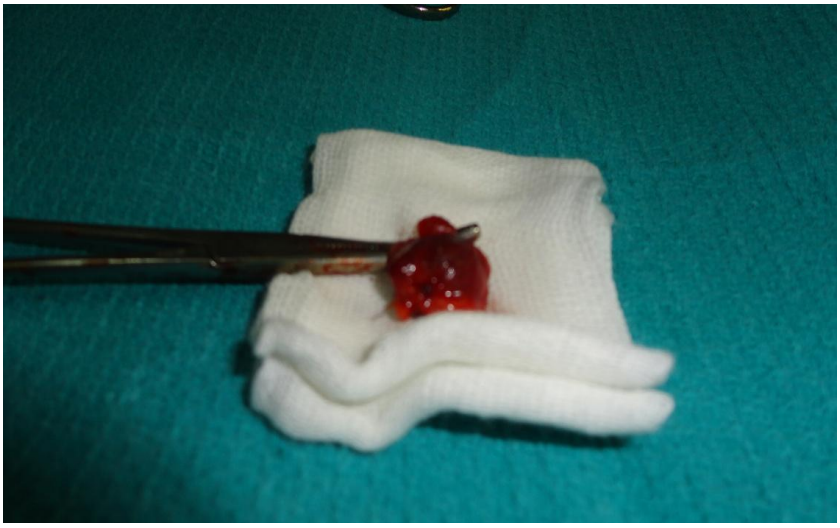

**Figure 1.3: shows the Excised specimen (single congested nodular tissue of measuring 1.5x1.3x0.8 cms)**

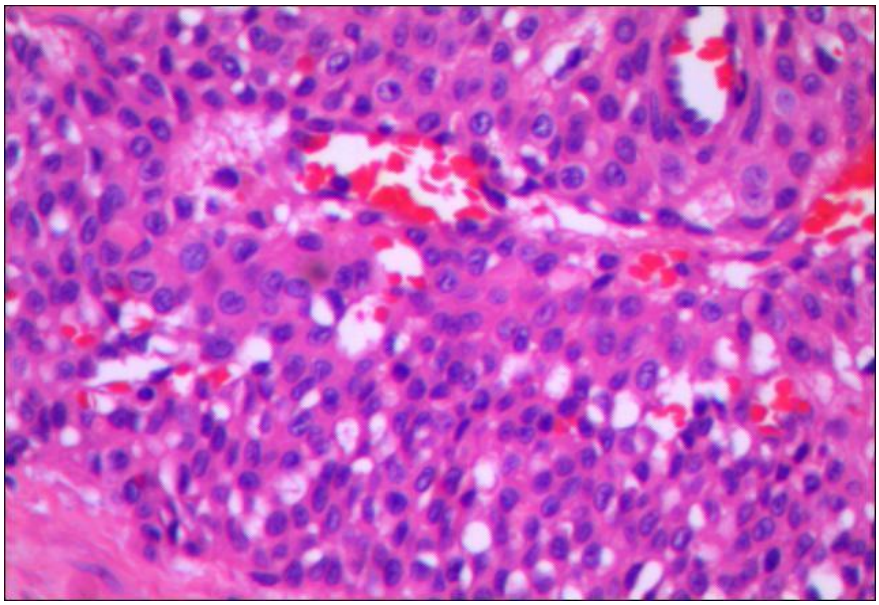

**Figure 1.4: Shows HPE was consistent with the features of Glomangioma**

## DISCUSSION

Glomus tumors are relatively rare vascular tumors with reported incidence of 1.6% of all soft tissue tumors and 4.5% of all tumors on the hand (Shugart *et al.*, 1963). It is variously regarded as a hamartoma or a neoplasm of neuromyoarterial glomus, which consists of dilated vascular channels, surrounded by proliferating glomus and nerve cells (Sucquet-Hoyer canal) and plays an important role in temperature regulation. However, as they are not of neoplastic nature, the term "glomangioma" was challenged by recent literature, authors considering that GVM represents the proper name. GVMs were described as early as the 18th century. However, it was William Wood in 1812 to first coin a name, specifically "painful subcutaneous tubercle," to describe the painful lesion treatable by surgical extirpation. In 1924, Pierre Masson, reported that the lesion described by Wood was a pathology of the dermal arteriovenous (AV) shunt (Smyth, 1971). Further development in understanding GVMs was made with the discovery of the Sucquet-Hoyer canal, which has a thick medial coat consisting of glomus cells (large, cuboidal cells) (Smyth, 1971). The Sucquet-Hoyer canal is an AV anastomosis with role in thermoregulation (Quigley, 1979). GVMs are commonly inherited and it is believed that they are caused by a mutation in the glomulin gene, which has a key role in the differentiation of vascular smooth-muscle cells (VSMCs), especially those belonging to cutaneous veins. Inherited GVMs have 197 been linked to chromosome 1p21-22 and are the result of truncating mutations in glomulin; these mutations have been found in all cases of familial GVM (Caughey and Highton, 1966). Most lesions occur in the dermis or subcutaneous tissue of the upper and lower extremities and have a diameter of less than 1 cm. Two forms have been described: the solitary form (90% of cases) and a multiple variant (10% of cases), the latter being more common in children and believed to have an autosomal dominant pattern of inheritance. The most common location for solitary, sporadic GVMs is the nail bed, which is characterized by the presence of numerous glomus bodies (Negri *et al.*, 1997). Approximately 80% of GVMs are affecting the upper extremities and 75% of these can be found subungally (Gencosmanoglu *et al.*, 2003). GVMs usually present as pink to purple nodular, hyperkeratotic lesions that are limited to the subcutaneous tissue and illicit pain when compressed (Weedon, 2002). Pain in GVMs correlates with the size of the lesion; the bigger the lesion, the more painful it is; in lesions greater than 5 cm, an approximate 80% of patients feel pain (Weedon, 2002). They are classified into 2 variants: solitary glomus tumors and multiple glomus tumors, with clinical and anatomopathologic differences. Solitary tumors are the most common and are the subject of the discussion in this article. They can manifest in the form of erythematous-violaceous nodule, red-pink or blue tipped macule, or increased curvature or deformity of the nail plate (Brouillard *et*

*al.,* 2002). Glomus tumors are classified according to their site as digital and extradigital. Extradigital tumors are less common. The typical clinical presentation is one of localized pain and sensitivity and hypersensitivity to cold.

**Table 1: Clinical Differences between Subungual and Extradigital Glomus Tumors (Brouillard *et al.*, 2002)**

| Clinical Characteristics | Subungual Glomus Tumor | Extradigital Glomus Tumor |
|---|---|---|
| Sex | Female 2:1 | Male 4:1 |
| Age on diagnosis | Twenties and fifties | Forties and sixties |
| Clinical characteristics | Pain, localized sensitivity and hypersensitivity to cold | Pain |
| Correct initial diagnosis | Often | Less often |

The commonest site is the hand, particularly the fingers. There have been reports in the literature of unusual location of glomus tumor such as ankle (Smyth, 1971), foot (Quigley, 1979), knee (Caughey and Highton, 1966), thigh (Negri *et al.,* 1997) and hip (Gencosmanoglu *et al.,* 2003). Glomus tumors may be solitary or multiple; the latter may be further divided into regional or disseminated, which are usually familial or congenital. Other variants such as plaque type and patch type have been described.

Histologically, the tumors have variable quantities of glomus cells, blood vessels and smooth muscles.Accordingly, they are classified as solid glomus tumors (25%), glomangioma (60%) and glomangiomyoma (15%) (Brouillard *et al.,* 2002). Solid glomus tumors are composed of aggregates of glomus cells surrounding inconspicuous vessels. Glomus cells are round, regular with pale or eosinophilic cytoplasm and dark staining round to oval nuclei. Glomangiomas have more prominent vessels and less conspicuous glomus cells.In glomangiomyoma there is an increase in number of spindle shaped smooth muscle cells which tend to be distributed near the vascular spaces and blend with adjacent glomus cells (Brouillard *et al.,* 2002). The histological features in our case were that of a glomangioma. Glomangioma has to be differentiated from the painful tumors of the skin such as eccrine spiradenoma, where two populations of cells and focal ductal differentiation are seen from leiomyoma and neuromatoid hyperplasia. Multiple hemangiomas have to be differentiated from blue rubber naevus.

Ultrasound has only recently been used as an additional study for diagnosis and follow-up of cutaneous lesions. Applications include a range of inflammatory diseases, benign and malignant tumors, and even esthetics (Boon *et al.,* 1999). In the ultrasound images captured in B mode, glomus tumors are seen as small solid, hypoechoic, well-defined nodules, more or less homogenous, with a regular border, most often located in the superficial dermis, with no involvement of the deep layers (Shivaswamy *et al.,* 2003). The color Doppler study shows extensive vascularization inside the tumor, as would be expected with a vascular type tumor such as a glomus tumor (Boon *et al.,* 1999). In addition, the pulsed Doppler study usually shows a low-grade systolic murmur. Both characteristics help us to differentiate such tumors, in the first instance, from other nonvascularized soft tissue tumors (Stout, 1937). However, an intense vascular signal in the Doppler study can also be seen in hemangiomas or arteriovenous malformations. Some B-mode ultrasound findings can assist in the differential diagnosis. In hemangiomas, grey-scale images do not show such homogenous echogenicity and the margins are less well defined. In addition, vascular flow is diminished as blood pools are present. Malformations are usually more poorly defined lesions compared to surrounding tissue, with no mass effect, and they show the typical arteriovenous shunt. Three of our patients had the typical stalk sign. This sign corresponds to a hypoechoic prolongation of the lesion by the stalk, which in Doppler mode reveals an intense vascularization characteristic of vascular lesions such as glomus tumors. Nevertheless, images obtained by ultrasound are not specific to any particular lesion, and diagnosis of glomus tumor is clinical, with confirmation from histopathologic study. However, cutaneous ultrasound can guide diagnosis, providing

significant information in a simple skin examination. Study of ultrasound images is also very useful for exactly, simply, quickly, and painlessly locating the lesion prior to excision. The technique can reveal the relationship with surrounding tissue to enable complete excision with minimal trauma and so avoid subsequent recurrence (Lendrum, 1947). Ultrasound has been able to detect tumors as small as 1 mm, reflecting its usefulness in the early diagnosis of lesions. The limitations of ultrasound are seen with flat lesions less than 3 mm across and with sites where artifacts may appear, such as subungual if the nail is very thick (Lendrum, 1947). The use of supplementary magnetic resonance imaging is a useful, noninvasive method, but it is more costly and so its use in everyday clinical practice does not appear to be justified. Radiography is inexpensive and quick, but it is only useful in large tumors and in subungual tumors to differentiate them from subungual exostosis.

The treatment of choice for isolated glomangioma is surgical excision. Sclerotherapy with sodium tetra decyl sulphate, polidocinol and hypertonic saline has been reported to be effective in patients with multiple glomangioma located on the extremities. However, sclerotherapy was found to be ineffective in a series of seven patients with large glomangioma. Ablative therapy with Argon and Carbondioxide laser is of potential benefit for small superficial lesions.

***Conclusion***

Misdiagnosis and/or delayed diagnosis of GVMs are common. Providing clinicians with guidelines for the proper identification and treatment of these conditions is crucial for the welfare of patients. Surgical excision is by far the most suitable method of treatment, especially for single GVMs, yielding excellent results. Once these lesions are properly diagnosed and treated, patients can experience an immediate improvement in the quality of their life.

## REFERENCES

**Boon LM, Brouillard P, Irrthum A, Karttunen L, Warman ML, Rudolph R, Mulliken JB, Olsen BR and Vikkula M (1999).** A gene for inherited cutaneous venous anomalies ("glomangiomas") localizes to chromosome 1p21-22. *The American Journal of Human Genetics* **65**(1) 125-33.

**Brouillard P, Boon LM, Mulliken JB, Enjolras O, Ghassibé M, Warman ML, Tan OT, Olsen BR and Vikkula M (2002).** Mutations in a Novel Factor, Glomulin, Are Responsible for Glomuvenous Malformations ("Glomangiomas"). *The American Journal of Human Genetics* **70**(4) 866-874.

**Brouillard P, Ghassibé M, Penington A, Boon LM, Dompmartin A, Temple IK, Cordisco M, Adams D, Piette F, Harper JI, Syed S, Boralevi F, Taïeb A, Danda S, Baselga E, Enjolras O, Mulliken JB and Vikkula M (2005).** Four common glomulin mutations cause two thirds of glomuvenous malformations ("familial glomangiomas"): evidence for a founder effect. *Journal of Medical Genetics* **42**(2) e13.

**Caughey DE and Highton TC (1966).** Glomus tumor of the knee: Report of a case. *Journal of Bone and Joint Surgery - British Volume* **48** 134–7.

**Chatterjee JS, Youssef AH, Brown RM and Nishikawa H (2005).** Congenital nodular multiple glomangioma: a case report. *Journal of Clinical Pathology* **58**(1) 102-3.

**Gencosmanoglu R, Inceoglu R and Kurtkaya-Yapicier O (2003).** Glomangioma of the hip. *Dermatologic Surgery* **29** 1244–7.

**Lendrum AC (1947).** Painful Tumours of the Skin: Post-Graduate Lecture given at The Royal College of Surgeons of England on 28th February, 1947. *Annals of the Royal College of Surgeons of England* **1**(2) 62-8.

**Negri G, Schulte M and Mohr W (1997).** Glomus tumor with diffuse infiltration of the quadriceps muscle: a case report. *Human Pathology* **28** 750–2

**Quigley JT (1979).** A glomus tumor of the heel pad: A case report. *Journal of Bone & Joint Surgery, American Volume* **61** 443–4.

**Shivaswamy KN, Thappa DM and Jayanthi S (2003).** A solitary painful nodule. *Indian Journal of Dermatology, Venereology and Leprology* **69** 359–60.

**Shugart RR, Soule EH and Johnson EW Jr (2003).** Glomus Tumor. *Surgery, Gynecology, and Obstetrics* **117** 334–40.
**Smyth M (1971).** Glomus-cell tumors in the lower extremity. Report of two cases. *Journal of Bone & Joint Surgery, American Volume* **53** 157–9.
**Stout AP (1937).** Solitary cutaneous and subcutaneous leiomyoma. *American Journal of Cancer* **29** 435-469
**Walsh JJ and Eady JL (2004).** Vascular tumors. *Hand Clinics* **20** 261–8.
**Weedon D (2002).** Vascular tumours. In: *Skin Pathology*, 2nd edition, edited by Weedon D (Churchill-Livingston, London) 1016–7.