



Case Report

Glomus Tumor: Comparative Analysis of Four Cases

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ABSTRACT

Glomus tumors are benign perivascular neoplasms that are uncommon and are rarely reported outside their usual peripheral soft tissue sites. Subungual region of the finger is the most common site, but other common sites include the palm, wrist, forearm and foot. In the current study we report four cases of glomus tumor at unusual sites. Histologically, glomus tumors are composed of uniform, round cells surrounding vascular spaces of varying sizes. On the basis of special stains and immunohistochemical characteristics; normal glomus cell is thought to be a modified smooth muscle cell origin. Majority of these tumors are benign; however, rare cases with atypical/malignant behavior have been reported ^[1-4]. The diagnosis of glomus tumor is finally confirmed on histopathology and it is cured by simple excision.

Keywords: Glomus, Vascular, Smooth Muscle.

INTRODUCTION

Glomus tumors consists of less than 2% of soft tissue tumors ^[5,6]. Most frequently occurring in the extremities, typically in the subungual region of the fingers. Rarely, they involve the internal organs such as mediastinum, lung, trachea, and stomach ^[7,8]. The tumor is about equally common in both genders, although there is a striking female predominance (3:1) among patients with subungual lesions. ^[9] These tumors are usually solitary, deep blue to purple in color, and accompanied by the symptoms of a triad of pain, cold sensitivity

and point tenderness ^[10]. The tumor is generally thought to arise from the arterial part of the glomus, the arteriovenous shunting system regulating blood flow in the distal extremities ^[11]. Extra digital glomus tumors are difficult to diagnose due to their non-specific clinical features, unusual sites and symptoms which vary compared with those of classical glomus tumors. Therefore, it is vital to keep in mind glomus tumors as a possible differential diagnosis in patients with extra digital lesions. In the present study four cases of glomus tumor are reported

involving the following sites; anterior chest wall, right knee, right thigh and left wrist.

MATERIALS AND METHODS

Four cases of glomus tumor were studied in the Department of Pathology, MGM Medical College

and Hospital, Kamothe, Navi Mumbai. The pertinent clinical data are given in Table 1. For histologic evaluation, hematoxylin - eosin, special stains; Periodic Acid Schiff (PAS) and Alcian Blue, ImmunoHisto Chemistry (IHC); Vimentin, CK and EMA were used in all cases.

Table no.1

Case No	Sex	Age (Years)	Site	Duration	Diagnosis
1	Female	62	Anterior Chest Wall	6 months	Glomus Tumor
2	Male	30	Right Knee	3 years	Glomangioma
3	Male	32	Right Thigh	1 year	Glomus Tumor
4	Male	21	Left Wrist	4 months	Glomus Tumor

Case No. 1

A 65 year old lady presented with a cystic, mobile swelling, not attached to the overlying skin at anterior chest wall since six months. Cystic mass was excised and sent for histopathological examination.

Case No. 2

A 30 year old male presented with single, localized, erythematous papule which was tender, extremely painful on Right Knee since three years. Dermatologist gave the differential diagnosis of Spiradenoma and Eruptive Yellow Cyst. Biopsy was taken and sent to histopathology department for final diagnosis.

Case No. 3

A 32 year old male presented with a mobile swelling on right thigh with bluish discoloration, solitary and tender since one year. The swelling was excised by the surgeon and sent for histopathology. Grossly, a single grey brown mass measuring 3.5x3x 1.2 cm was received. On the summit of the mass a single blue nodule measuring 0.5 cm was noticed.

Case No. 4

A 21 year old male presented with a slow growing mass over left wrist since 3- 4 months after a history of trauma. Grossly, single, soft to firm, grey brown, skin covered tissue bit measuring 0.5 cm was received which was further processed for histopathological examination.

RESULTS

In all the cases tumor cells were arranged into multiple, circumscribed lobules, anastomosing cords or collar-like structures with well-defined cell borders along with abundant small vessels between them (Fig. 1,2 &3). Individual tumor cells were uniform, round to oval with absent or minimal nuclear atypia, vesicular chromatin and occasional single prominent nucleolus (Fig. 4). The cytoplasm was relatively dense eosinophilic and abundant. Stroma was loose, myxoid containing numerous vessels of varying size, some with hemangiopericytoma-like configuration. There was no evidence of spindle cells, tumor necrosis, and intravascular invasion in the sections studied. The mitotic rate varied from 1 to 3 per 50 high-powered fields (HPFs).

Sections stained with special stains; PAS and Alcian blue gave positive results (Fig.5).

Immunohistochemically, the tumor cells were immunoreactive for vimentin (Fig. 6) and were negative for cytokeratin and EMA.

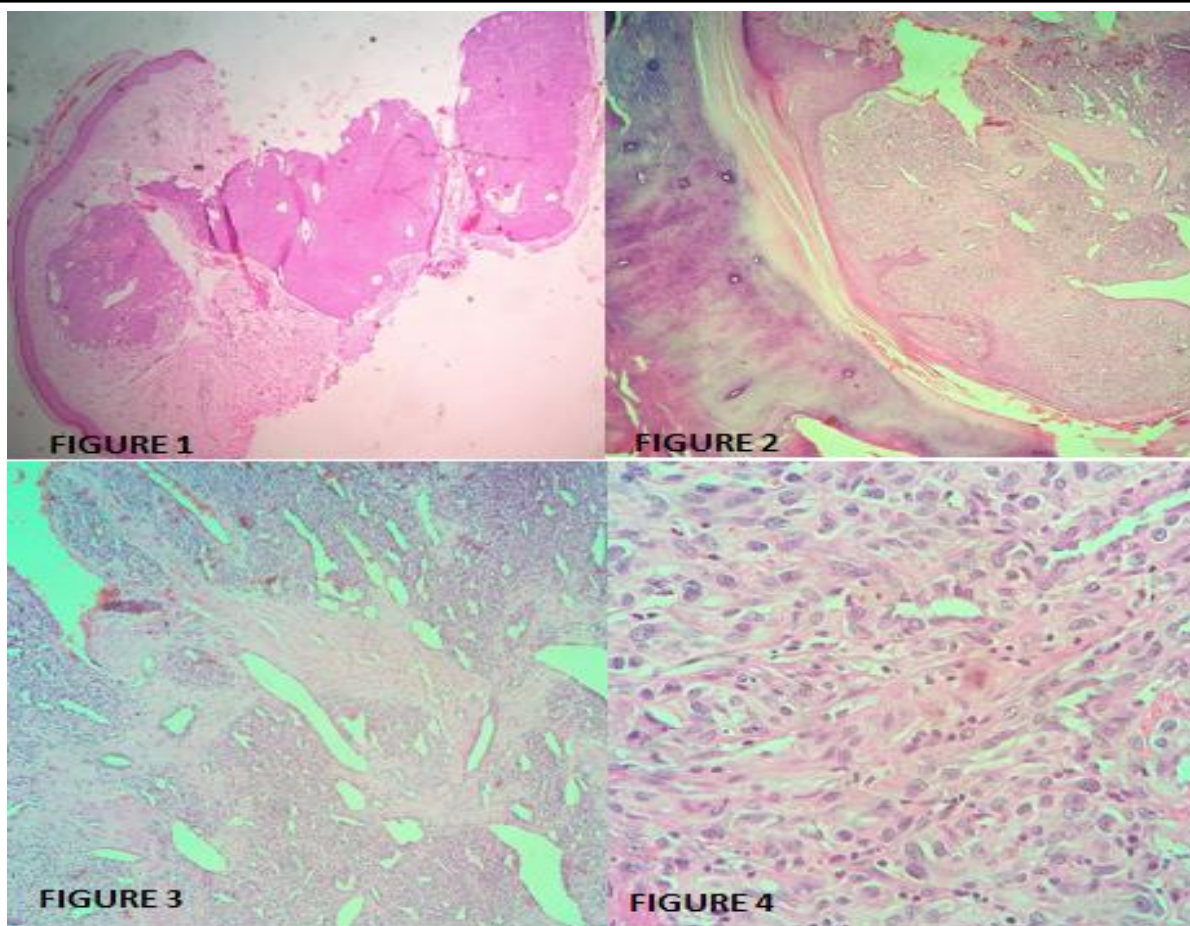


Figure 1 & 2 (40 X, H&E stain) shows tumor cells in multiple, circumscribed lobules.

Figure 3 (100 X, H&E stain) shows perivascular arrangement of tumor cells.

Figure 4 (400 X, H&E stain) shows uniform round to oval tumor cells.

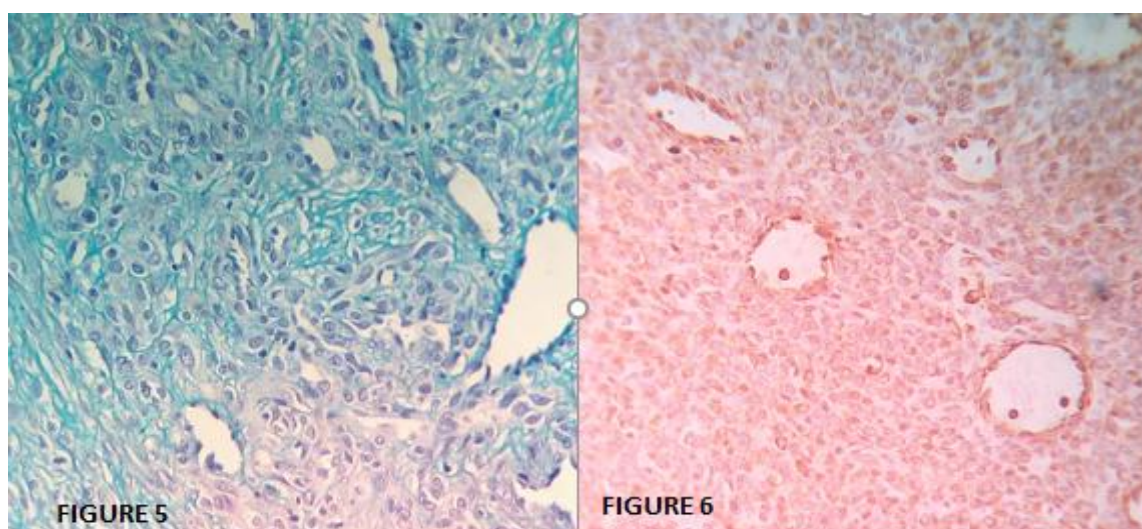


Figure 5 (400 X, Alcian Blue stain) shows positive tumor cells.

Figure 6 (400 X, Vimentin ;IHC stain) shows positivity in the tumor cells.

DISCUSSION

Glomus tumor was first described in 1924 by Masson ^[12], as a distinct perivascular neoplasm that is believed to originate from modified smooth

muscle cells that are present in the walls of specialized arteriovenous shunts (Sucquet-Hoyer canals) engaged in thermoregulation. The tumor is about equally common in both genders, although

there is a striking female predominance (3:1) among patients with subungual lesions. ^[9]. Beaton *et al* ^[13] suggested that the frequency of extra digital cases varied from 11 to 65% and may be more common in males than females. Lee *et al* demonstrated that extra digital glomus tumors are more common in males, whereas digital tumors are more frequent in females ^[10]. As seen in our study all the cases were present at extra digital sites and out of the four cases 3 were males.

Clinically, glomus tumors are characterized by a triad of sensitivity to cold, localized tenderness and severe and intermittent pain. ^[14] The pain is excruciating and burning or bursting in nature which is believed to be because of nerve fibers containing the pain neurotransmitter substance P have been involved in the tumor. ^[15] Glomus tumors are well circumscribed and composed of varying proportions of glomocytes, blood vessels and smooth muscle. On the basis of proportions of the components, glomus tumors are divided in three subgroups: glomus tumor proper, glomangioma, and glomangiomyoma. Glomus tumors are usually benign; however, rare cases of atypical and malignant glomus tumors with recurrences, metastases, and death have been reported ^[1-4]. In our study all the cases were diagnosed as benign glomus tumor except one which was diagnosed as glomangioma due to predominance of blood vessel component.

The main differential diagnostic consideration for glomus tumor could be other perivascular tumors such as hemangiopericytoma, juxtaglomerular tumor; smooth muscle neoplasms such as epithelioid leiomyoma, carcinoid tumor, and paraganglioma. Glomus tumors can have a hemangiopericytomatous (staghorn) vascular pattern. We can distinguish them on the basis of IHC; hemangiopericytomas are negative for actin and positive for CD34 and lack the uniform round cells that are seen in glomus tumors. ^[16]

Immunohistochemical analysis, on the other hand, indicated that the glomus tumor cells contained intermediate filaments of the vimentin type, but

desmin, normally found in smooth muscle cells ^[17,18,19] could not be detected. The lack of desmin does not exclude the possibility of a smooth muscle-derivation, since all smooth muscle cells do not contain desmin and may express vimentin instead ^[17]. For example, aortic smooth muscle cells in the proximal aorta between the large vessels and the bifurcation, seem to contain only vimentin ^[17,20]. In the present study conducted only vimentin was positive.

Accurate diagnosis followed by complete excision is the treatment of choice for patients with solitary lesions, and recurrence rates for solitary tumors are 12 to 33% ^[21,22]. It is rare for malignant glomus tumors to occur.

In conclusion, we reported four cases of extra digital glomus tumor arising in anterior chest wall, right knee, right thigh and left wrist. Varying clinical symptoms from classical glomus tumor and presence of tumor at unusual sites occasionally interfere with the diagnosis and treatment of patients. Therefore, it is important to include the glomus tumor in the differential diagnosis of patients with extradigital painful or asymptomatic lesions that are purple in color.

REFERENCES

1. Folpe AL, Fanburg-Smith JC, Miettinen M, et al. Atypical and malignant glomus tumors: analysis of 52 cases, with a proposal for the reclassification of glomus tumors. *Am J Surg Pathol* 2001;25:1 - 12.
2. Leu HJ. Glomus tumours: morbid-anatomical analysis of 58 cases, especially infiltrated forms [in German]. *Dtsch Med Wochenschr* 1981;106:171 - 4.
3. Gould EW, Manivel JC, Albores-Saavedra J, et al. Locally infiltrative glomus tumors and glomangiosarcomas. A clinical, ultrastructural, and immunohistochemical study. *Cancer* 1990;65: 310 - 8.
4. Watanabe K, Hoshi N, Tsu-Ura Y, et al. A case of glomangiosarcoma. *Fukushima J Med Sci* 1995;41:71 - 7.

5. Enzinger FM, Weiss SW. Perivascular tumors. In: Enzinger FM, Weiss SW, editors. Enzinger and Weiss' soft tissue tumors. 4th ed. St Louis (MO) Mosby; 2001. p. 985 - 1035.
6. Gaertner EM, Steinberg DM, Huber M, et al. Pulmonary and mediastinal glomus tumors—report of five cases including a pulmonary glomangiosarcoma: a clinicopathologic study with literature review. *Am J SurgPathol* 2000;24:1105 - 14.
7. Miettinen M, Paal E, Lasota J, et al. Gastrointestinal glomus tumors: a clinicopathologic, immunohistochemical, and molecular genetic study of 32 cases. *Am J SurgPathol* 2002;26:301 - 11. Fig. 2 (A) Strong cytoplasmic staining for calponin. (B) Type IV collagen immunostain with membranous staining outlining each tumor cell. Glomus tumor of renal pelvis 301
8. Sonobe H, Ro JY, Ramos M, et al. Glomus tumor of the female external genitalia: a report of two cases. *Int J GynecolPathol* 1994; 13:359 – 64.
9. Shugart RR, Soule EH, Johnson EW. Glomus tumor. *SurgGynecolObstet* 1963; 117:334.
10. Lee DW, Yang JH, Chang S, Won CH, Lee MW, Choi JH, Moon KC. Clinical and pathological characteristics of extradigital and digital glomus tumors: a retrospective comparative study. *J Eur Acad Dermatol Venereol*. 2011;25:1392–1397. [[PubMed](#)]
11. Lever WF (1975) Histopathology of the skin. Lippincott, Philadelphia, pp 600-603 Miettinen M, Lehto V-P, Badley RA, Virtanen I (1982) Expression of intermediate filaments in soft tissue sarcomas. *Int J Cancer* 30:541-546.
12. Masson P. Le glomus neuromyoarterial des regions tactiles et sesteumeures. *Lyon Chir* 1924;2:257 – 80.
13. Beaton LI, Davis L. Glomus tumor: report of three cases and analysis of 271 recorded cases. *Q Bull Northwest Univ Med School*. 1941;15:245–273.
14. Vandevender DK, Daley RA. Benign and malignant vascular tumors of the upper extremity. *Hand Clin* 1995; 11: 161-79.
15. Kishimoto S, Nagatani H, Miyashita A, Kobayashi K. Immunohistochemical demonstration of substance P containing nerve fibers in glomus tumors. *Br J Dermatol* 1985; 113: 213.
16. Rosai J. Urinary tract. In: Rosai J, editor. Rosai and Ackerman's Surgical Pathology. 9th ed. 2004. p. 1163 – 317
17. Frank ED, Warren L (1981) Aortic smooth muscle cells contain vimentin instead of desmin. *Proc Natl Acad Sci USA* 78: 3020-3024
18. Gabbiani G, Schmid E, Winter S, Chaponnier C, de Chastonay C, Vandekerckhove J, Weber K, Franke W (1981) Vascular smooth muscle cells differ from other smooth muscle cells: Predominance of vimentin filaments and a specific-type actin. *Proc Natl Acad Sci USA* 78: 298-30
19. Miettinen M, Lehto V-P, Badley RA, Virtanen I (1982) Expression of intermediate filaments in soft tissue sarcomas. *Int J Cancer* 30:541-546
20. Osborn M, Caselitz J, Weber K (1981) Heterogeneity of intermediate filament expression in vascular smooth muscle: A gradient in desmin positive cells from the rat aortic arch to the level of the arterial iliacacommunis. *Differentiation* 20:196-202.
21. Rettig AC, Strickland JW. Glomus tumor of the digits. *J Hand Surg Am*. 1977;2:261–5. [[PubMed](#)]
22. Strahan J, Bailie HW. Glomus tumour. A review of 15 clinical cases. *Br J Surg*. 1972;59:91–3. [[PubMed](#)]