Symplastic Glomus Tumor: Pathologist's Challenge and Physician's Dilemma: A Case Report with Review

Abstract

Glomus tumor is a distinct mesenchymal tumor of specialized perivascular smooth cell origin (pericyte), reminiscent of normal glomus body located in the fingers. It accounts for <2% of soft tissue tumors. Recently WHO categorized few unusual variants, of which symplastic glomus tumor is one. Histologic picture of the latter shows marked nuclear atypia in the absence of any other malignant features. They are exceedingly rare, hence their biological behavior, prognosis and treatment are little understood. But recognizing this entity would prevent misdiagnosis of malignancy. We report such a case in a 39-year-old female presented with excruciating pain in the left thumb.

Keywords: *Glomus tumor, malignant potential, symplastic*

Pranita Mohanty, Ajit S. Mohapatra, Ritu P. Choudhury, Maitreyee Panda[1]

Departments of Pathology and [1]Skin and Veneral Diseases, IMS and SUM Hospital, S'O'A (Deemed to be) University, Bhubaneswar, Odisha, India

Introduction

Glomus tumors are uncommon benign mesenchymal neoplasms accounting for <2% of the soft tissue tumor.[1] It arises from specialized cells (pericytes) that resemble glomus apparatus in the skin, involved in thermoregulation. Glomus tumor was first described by Wood (1812) and the histological description was given by Barre and Masson (1924).[2-4] WHO categorized these tumors under perivascular (pericytes) soft tissue tumor with many variants.[1] They commonly present as "painful subcutaneous nodule" characterized by a triad of spontaneous pain, tenderness, and cold sensitivity. The usual cutaneous forms are subcentrimetric (<2 cm) solitary dermal nodule, predominantly located in subungual region of distal phalanx with a reddish purple spot over the nail. However, extracutaneous sites' occurrence such as bone, nerve, penis, mediastinum, liver, stomach, colon, kidney, trachea, and lungs has also been published in literature.[1,4] Glomus tumors are most common in 4th–6th decade. Any sex can be affected equally except female preponderance seen in subungual locations. Histopathological examination of a typical glomus tumor is characterized by a well-circumscribed dermal lesion, consisting of tight convolutes of capillary-sized vessels, surrounded by collars of glomus cells, set in a hyalinized or myxoid stroma. Glomus cells are uniform small rounded having regular margins, eosinophilic cytoplasm and darkly staining round to oval nuclei. Immunohistochemical techniques demonstrates positivity for vimentin, muscle-specific actin (MSA), α-smooth muscle actin (α SMA), type IV collagen, H-caldesmon, and low Ki 67 index. Presentation of this case was aimed at making aware of this rare entity and prevent misdiagnosis of malignancy with inadvertent overtreatment.

Case Report

A 39-year-old female presented to dermatology department with sudden increase in pain in the left thumb since five days following gradual pain over a period of four years for which she underwent treatment from local hospital that had given only symptomatic relief. There was no history of trauma. Clinical examination revealed a minute reddish blue spot beneath the nail bed of the left thumb. Her contrast-enhanced MRI of the left thumb showed an oval T2W hyperintense lesion measuring (1.9 × 7.2 × 5.9 mm) in extensor aspect of distal phalanx/nail bed of the thumb with intense homogenous post-contrast enhancement. Impression of suspected glomus tumor was given. Surgical excision was done and submitted for histopathology.

Address for correspondence:
Dr. Pranita Mohanty, Professor, Department of Pathology, IMS and SUM Hospital, Bhubaneswar - 751 003, Odisha, India.
E-mail: dr.pranitamohanty@gmail.com

Access this article online

Website: https://journals.lww.com/idoj

DOI: 10.4103/idoj.idoj_507_21

Quick Response Code:

How to cite this article: Mohanty P, Mohapatra AS, Choudhury RP, Panda M. Symplastic glomus tumor: Pathologist's challenge and physician's dilemma: A case report with review. Indian Dermatol Online J 2023;14:682-5.

Received: 09-Aug-2021. **Revised:** 24-Apr-2023. **Accepted:** 19-May-2023. **Published:** 15-Aug-2023.

Microscopy revealed a well-circumscribed oval lesion showing solid proliferation of round glomus cells having distinct margin, round nuclei and eosinophilic cytoplasm seen in a myxoid background surrounding many convoluted capillaries [Figure 1]. Good number of cells exhibited prominent cytological atypia with marked nuclear enlargement and pleomorphism, anisonucleosis, hyperchromasia, pseudoinclusions, bizarre nuclei, coarse chromatin, and irregular nuclear membrane [Figure 2]. However, no mitoses/atypical mitotic figure or necrosis was identified. Immunohistochemistry (IHC) for vimentin and SMA showed both cytoplasmic and membranous positive with ki-67 index =3% [Figures 3 and 4]. CD34 highlighted vessel wall as well as tumor cells [Figure 5], whereas S100 was negative in tumor cells [Figure 6]. Final diagnosis of symplastic glomus tumor was made.

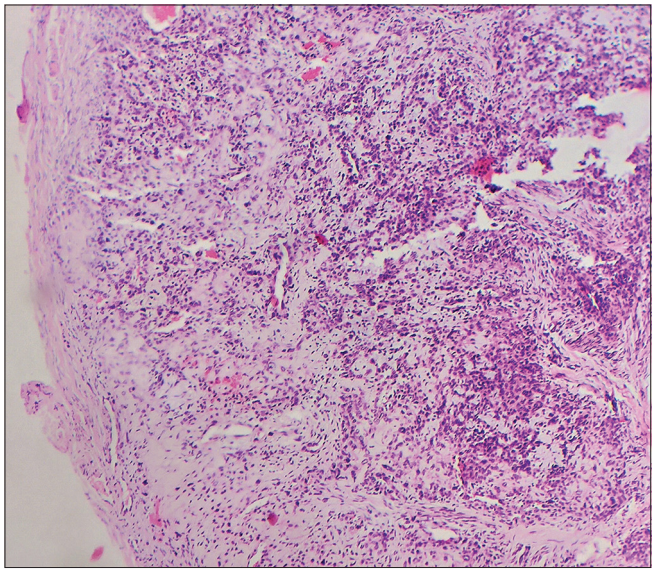

Figure 1: Photomicrograph showing solid sheets of fairly monomorphic cells surrounding capillaries (H and E, 10×)

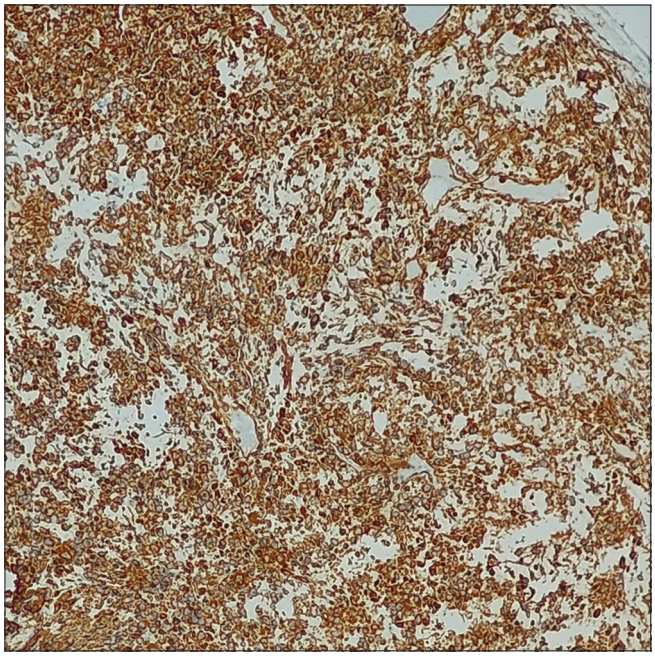

Figure 3: Photomicrograph showing vimentin positivity (10×)

Discussion

The first report of a clinically atypical infiltrating glomus tumor was by Lumley and Stansfeld.[5] One year later, the 40th Assembly of the Greek Pathological Society and Anagnostou *et al*.[6] presented individual cases of pathologically malignant glomus tumor. Folpe *et al*.[7] proposed for re-classification scheme and criteria with a study comprising 52 unusual glomus tumor cases, of

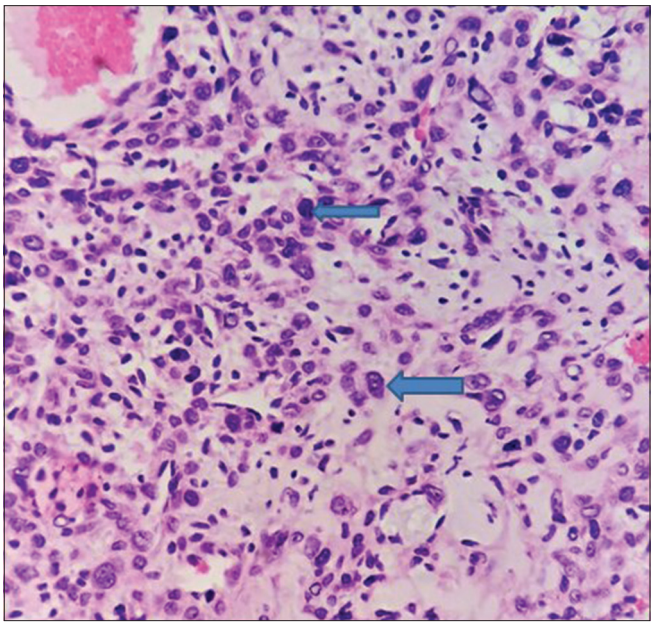

Figure 2: Photomicrograph showing prominent cytological atypia with marked nuclear enlargement and pleomorphism, anisonucleosis, hyperchromasia, pseudoinclusions, bizarre nuclei, coarse chromatin, and irregular nuclear membrane (blue arrowed) (H and E, 40×)

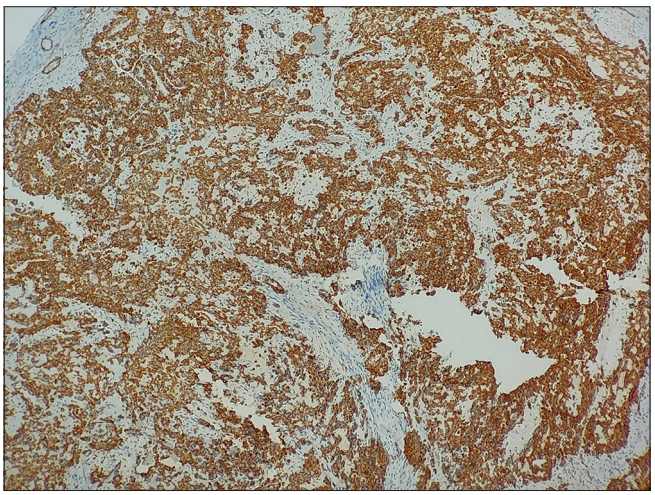

Figure 4: Photomicrograph showing smooth muscle actin (SMA) positivity (10×)

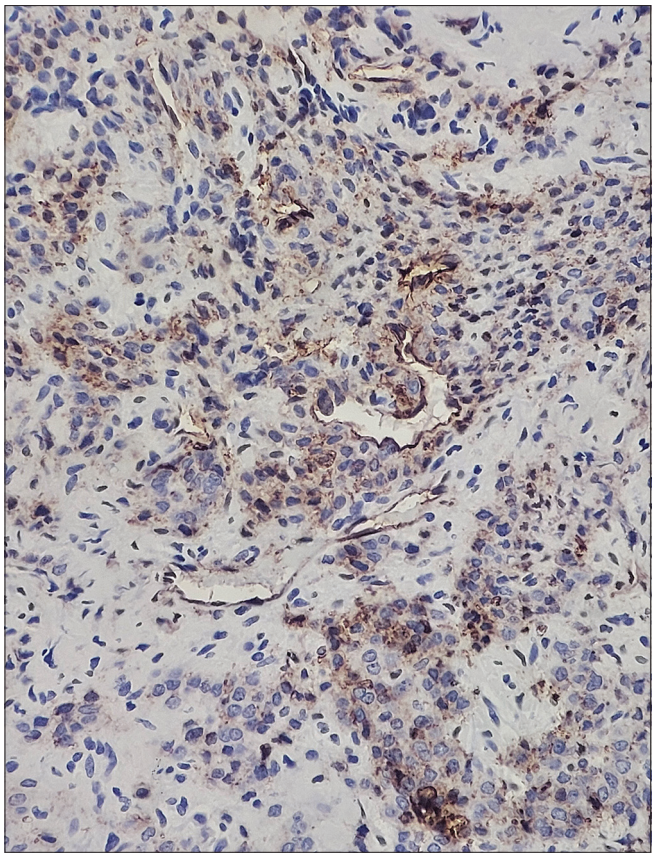

Figure 5: Photomicrograph showing CD 34 positivity (10×)

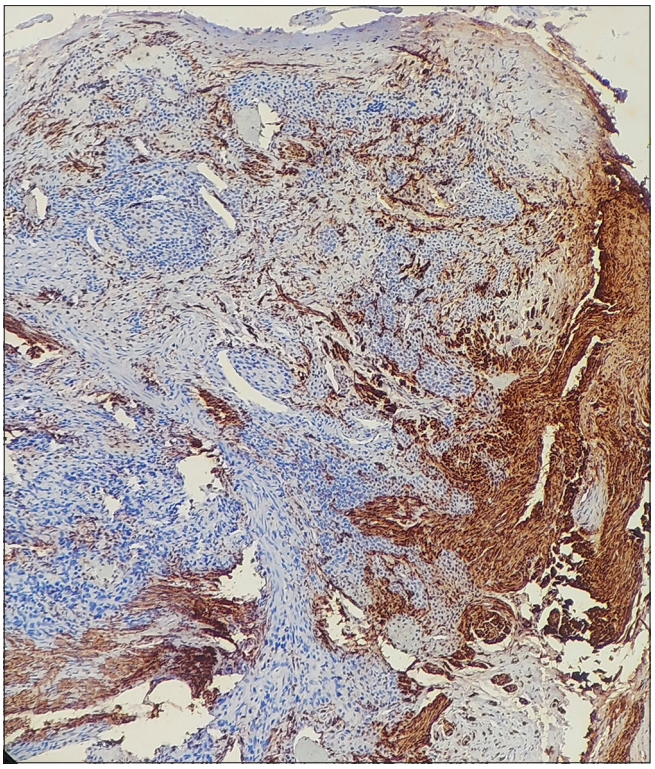

Figure 6: Photomicrograph showing S100 negativity (10×)

which nine cases were catagorized as symplastic glomus tumors and disclosed two recurrences and no metastases with average of ten years follow-up information in six of those nine patients. They concluded that these tumors have a benign course, similar to ordinary glomus tumors. The diagnostic malignancy criteria suggested were: tumors with a deep location and a size of more than 2 cm, or atypical mitotic figures, or moderate to high nuclear grade and >5 mitotic figures/50 hpf. But mitotic activity of >5 / 50 hpf, high cellularity, presence of necrosis, and moderate-to-high nuclear grade did not show any significant relationship with metastasis. Based on their results, a new classification was done that included: (1) malignant glomus tumors, (2) symplastic glomus tumors, (3) glomus tumors of uncertain malignant potential, and (4) glomangiomatosis. In 2002, WHO, classified, published and reprinted in 2006 catagorizing them into 4 types: (1) Typical glomus tumor, (2) glomangiomatosis, (3) symplastic glomus tumor, and (4) malignant glomus tumor (glomangiosarcomas) and glomus tumors of uncertain malignant potential. Typical glomus tumors are subcategorized as "solid glomus tumor," "glomangioma," and "glomangiomyoma" depending on the relative prominence of glomus cells, vascular structures, and smooth muscle. The WHO criteria for "malignant glomus tumor" are: (1) size >2 cm and subfascial or visceral location; (2) atypical mitotic figures; or (3) marked nuclear atypia and any level of mitotic activity. Glomus tumors not fulfilling criteria for malignancy, but having at least one atypical feature other than nuclear pleomorphism, diagnosed as "glomus tumours of uncertain malignant potential." Symplastic glomus tumors are characterized by striking nuclear atypia/pleomorphism in the absence of any other worrisome malignancy feature and behaved in a benign fashion.[1,8] The marked nuclear atypia is believed to be a degenerative/senescence phenomenon. The term "symplastic" is coined to denote these nuclear features with homologous terminology like symplastic leiomyoma, symplastic hemangioma, ancient (not as symplastic) schwannomas.[4,9] Some authors suggested that nuclear atypia may be due to the accumulation of heterochromatin, associated with DNA inactivation.[9] Symplasic glomus tumor histomorphologically shows round cell morphology though not as monomorphic as that of typical glomus due to nuclear atypia and pleomorphism. But symplastic leiomyoma or ancient schwannomas are predominantly spindle cell tumors and have distinct histomorphology like long intersecting fascicles are seen in former, whereas zonations called Antoni A and Antoni B are observed in the latter. IHC of S100 (neural marker) show positivity for ancient schwannomas; vimentin and SMA (smooth muscle actin) positivity for symplastic leiomyoma/symplasic glomus. Usually, the location, clinical presentation, distinct histomorphology, and CD34 IHC distinguish both the entities.

Simillarly Lee *et al*.[10], Kim *et al*.[11], Chong *et al*.[12], Kabukçuoğlu F *et al*.[13], Da Silva *et al*.[14] described one case of symplastic glomus tumor each in subungual

Table 1: Reported symplastic glomus tumor in literatures

Author	Year of publication	Age (in years)	Sex (M/F)	Location
Arsenovic *et al*.[9]	2011	*NA	*NA	*NA
Kamarashev *et al*.[4]	2009	78	F	Nail deformity of the index finger of the left hand
Lee *et al*.[10]	2003	*NA	*NA	*NA
Kim *et al*.[11]	2005	*NA	*NA	*NA
Chong *et al*.[12]	2009	44	F	On the right index finger tip
Kabukçuoğlu *et al*.[13]	2015	37	F	The fourth finger tip of her left hand
Da Silva *et al*.[14]	2018	41	F	The left thumbnail
Falleti *et al*.[15]	2012	62	F	Right index finger

*NA – not available

location, followed up without post-surgery recurrence [Table 1]. Falleti *et al*.[15] reported one such case with six months follow-up and compared with a review of 14 more such cases till their study.

Commonest differentials are angioma, lipoma, and cyst. As tumor usually localized to the nail matrix without extension down the nail bed, the preferred treatment is the complete meticulous surgical excision with different approaches, such as dorsal trans-ungual/lateral sub subperiosteal/ Keiper–Litter approach to prevent recurrences.

Conclusion

Disease awareness among primary care physicians, proper diagnosis by pathologists, and possible treatment and outcome of this rare entity make it challenging yet curable. Hence it should be highlighted to avoid misdiagnosis of malignancy and to improve care of the patients without overtreatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Fletcher CDM, Unni KK, Mertens F. WHO classification of tumors. In: Pathology and Genetics of Tumors of Soft Tissue and Bone. Lyon: IARC Press; 2002.
2. Wood W. On painful subcutaneous tubercle. Edinburgh Med J Surg 1812;8:283-91.
3. Fletcher CDM, Bridge JA, Hogendoorn P, Mertens F. WHO Classification of Tumours of Soft Tissue and Bone. 4th ed. WHO; 2013. IARC WHO Classification of Tumours, No 5.
4. Kamarashev J, French LE, Dummer R, Kerl K. Symplastic glomus tumor – A rare but distinct benign histological variant with analogy to other ancient benign skin neoplasms. J Cutan Pathol 2009;36:1099-102.
5. Lumley JS, Stansfeld AG. Infiltrating glomus tumour of lower limb. Br Med J 1972;1:484-5.
6. Anagnostou GD, Papademetriou DG, Toumazani MN. Subcutaneous glomus tumors. Surg Gynecol Obstet 1973;136:945-50.
7. Folpe AL, Fanburg-Smith JC, Miettinen M, Weiss SW. Atypical and malignant glomus tumors: Analysis of 52 cases, with a proposal for the reclassification of glomus tumor. Am J Surg Pathol 2001;25:1-12.
8. Patterson JW, Strutton G, Rubin AI, Weedon D, ClinicalKey. Weedon's Skin Pathology. 5th ed. [Edinburgh]: Churchill Livingstone/Elsevier; 2021.
9. Arsenovic N, Ramaiya A, Moreira R. Symplastic glomangioma: Information review and addition of a new case. Int J Surg Pathol 2011;19:499-501.
10. Lee WC, Jang YH, Kim HJ, Lee SJ, Chung SL, Lee SK, *et al*. A case of symplastic glomus tumor. Korean J Dermatol 2003;41:1118-20.
11. Kim MJ, Lee NY, Song WK, Hahm JH, Whang KK. A case of a symplastic glomus tumor. Korean J Dermatol 2005;43:1301-4.
12. Chong Y, Eom M, Min HJ, Kim S, Chung YK, Lee KG. Symplastic glomus tumor: A case report. Am J Dermatopathol 2009;31:71-3.
13. Kabukçuoğlu F, Özkayalar H, Sakız D, Yavuz Kabukçuoğlu Y. Symplastic glomus tumor. Turk Patoloji Derg 2015;31:211-4.
14. Da Silva DR, Gaddis KJ, Hess S, Rubin AI. Nail unit glomus tumor with myxoid and symplastic change presenting with longitudinal erythronychia. Dermatopathology 2018;5:74-8.
15. Falleti J, Vita G, De Cecio R, Schonauer F, Insabato L, Natella V, *et al*. Symplastic glomus tumor: Report of a challenging lesion with literature review. Pathol Res Pract 2012;208:372-5.