

Glomus Tumour- Knowledge Update and Issues

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Dear Editor,

At the outset, I would like to congratulate the authors of the article entitled

“Giant Glomus Tumour of the Forearm- An Unusual Occurrence” published in July’2020 issue of JCDR (Vol-14(7):RD01-RD03) and DOI: 10.7860/JCDR/2020/43732.13868 [1], which I read with great interest. However, there are certain issues which need to be addressed for the benefit of the readers of this article with associated awareness of certain important information.

Glomus tumours are thought to arise from the glomus body or Sucquet-Hoyer canal, which works as thermoregulatory arteriovenous shunt composed of modified smooth muscle cells. Forearm as location of Glomus tumours is quite common and not an unusual occurrence as mentioned here [2,3]. Glomus tumours are sized as small tumours and large tumours which are more than 2 cm in size, however, literature does not mention anything with regard to size to qualify for “Giant” term as mentioned here by author. A size based terminology can be used only when there is case series of multiple cases for establishing such differentiation. In fact, it falls in the category of “Atypical glomus tumours”, where Folpe has included the glomus tumours with size more than 2 cm [4]. If there is no underlying inherited condition then the tumour is considered “sporadic” or “random”. Family history of glomus tumour should be taken to establish the inherited or sporadic form of the tumour, which the author has not specified.

It is important to mention for readers that Glomuvenous Malformations (GVMs), formerly known as glomangiomas, have also been described. While once considered a subset of glomus tumours, GVMs are now widely considered to be unrelated in pathogenesis given their differences in clinical and histopathologic features. It is thought that GVMs and glomus tumours have different aetiologies, with GVMs resembling venous malformations and containing more dilated venous channels than glomus tumours. World Health Organisation (WHO) who has classified glomus tumours based on their predominance of glomus cells, vascular structures, and smooth muscle cells as solid glomus tumours (most common, glomus cell prominence), glomangioma (vascular cell prominence), and glomangiomyoma (vascular and smooth muscle cell prominence). Besides, glomus tumour is classified based on continuum of severity as glomus tumour proper, glomangioma, and glomangiosarcoma [5-8]. Clinically, it is pertinent to mention about cold sensitivity test, which the author has not conducted and is a very easy test and is considered positive when immersing the affected area in cold water elicits severe pain around the lesion, a test even done by a medical practitioner without remembering other tests (like Love and Hildreth test as mentioned in the article), will in most cases can avoid missing the diagnosis of glomus tumour. While the vast majority of glomus tumours are benign, malignant cases have been rarely reported, with such cases typically being locally invasive, which is apparent in the case presented here by the author [1]. Although, malignant glomus tumours are more likely to be deep, larger than 2 cm, and have atypical features, metastases are exceedingly rare [9]. So, a tumour of such a large dimension as presented by author, should have thorough systemic evaluation to rule

out metastasis and also intravascular spread of the glomus tumour, which although rare has been described in the glomus tumour of stomach and subcutaneous tissue [9-13].

Systemic effects of glomus tumours are rare; however thrombocytopenia has been reported in the literature as a result of platelet sequestration. In these patients, a complete blood cell count is indicated, including platelet count [14]. Extracutaneous sites have been reported, including involvement of the gastrointestinal tract, trachea, nerve, bone, mediastinum, liver, pancreas, kidney, and ovary which the author has not elaborated and readers should be made aware of it too [15-19]. A consideration of the following differential diagnoses should be undertaken while evaluating for such lesions which includes-Blue Nevi, Blue Rubber Bleb Nevus Syndrome, Dermatologic Manifestations of Kaposi Sarcoma, Dermatologic Manifestations of Neurilemmoma (Schwannoma), Eccrine Spiradenoma, Fibroma, Haemangioma, Leiomyoma, Maffucci Syndrome, Mucoid cyst and Venous Malformations.

Immunohistochemistry in glomus tumour typically shows the tumour cells revealing immunopositivity for vimentin, smooth muscle actin and muscle specific actin in the cytoplasm. The cells do not stain with endothelial markers. Desmin is occasionally only focally positive. Moreover, vimentin positivity is found in 100% glomus tumours and adds on to the confirmation of glomus tumour which is reported here to be negative, questions the diagnosis of Glomus tumour proper [2,20]. Also, it should be noted that the neoplastic cells in glomus tumour may show a co-expression of alpha-smooth muscle actin and CD34, an important finding regarding the differential diagnosis of these lesions and the relationship of perivascular neoplasms [21,22].

It is also pertinent to mention that though, the histology of glomus tumour is quite characteristic but the cytologic features are poorly defined. The cytomorphologic picture may mimic that of an epithelial neoplasm, carcinoid tumour or a haemangiopericytoma and hence, may cause diagnostic difficulties and should be ruled out by immunohistochemistry [20,23-25]. Even though cytologic features of glomus tumour are similar to that of epithelial cells, meticulous attention is required with regard to clinical history and cytopathology. Finding of endothelial cells in close association to tumour cells indicates correct diagnosis. Sometimes, histologic sections of solid glomus tumour can masquerade as adnexal tumours like eccrine spiradenoma and hidradenoma. Immunohistochemical profile is very helpful in establishing the diagnosis. Glomus tumours are usually positive for vimentin and SMA, and negative for desmin, S-100, factor-VIII and epithelial markers [20].

In the case of solitary glomus tumours, complete removal of the tumour capsule is recommended to relieve pain and minimise risk for recurrence. However, modalities like laser and sclerotherapy have been reported, which should be mentioned to complete the information with regard to readers of this article.

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