Case Report

Case report of middle aged male patient with extra digital glomus tumor visiting dermatology clinic of King Abdul-Aziz Medical City, National Guard Health Affairs, Jeddah, Saudi Arabia, 2018

Awadh Alamri¹, Marwan Alahmadi², Mohamed B. Satti³, Amaal F. Alruwaili⁴, Homaid Alotaibi⁵, Hani Saad Al-Mugti⁶*

INTRODUCTION

The normal glomus body is located in the lower layer of the dermis (or stratum reticulare) throughout the body, but is more concentrated in the digits. Physiological function of the glomus body is a thermal regulation by shunting the blood away from the skin surface when exposed to cold temperature, thus preventing heat loss, and allowing maximum heat flow to the skin in warm weather to allow heat to dissipate.¹

Glomus tumors are a rare benign neoplasm arising from the glomus body and mainly found under the nail, on the fingertip or in the foot. The average age at presentation is from 30 to 50 years of age, although can occur at any age. Usually misdiagnosis with other skin tumor and unfortunately correct diagnosis takes seven years from onset of symptoms.²

There are multiple factors contribute to delay the diagnosis of this kind of tumor, especially for locations other than digital site, including low clinical suspicion,
variety of sites that can be affected and the different symptoms that patients may present with. Glomus tumor of the forearm is an example of uncommon location of the tumor, which differs slightly in its presentation.

In this article, the researchers decided that they should report a case of glomus tumor in the forearm and compare it to digital tumors in term of symptoms and diagnostic methods.

CASE REPORT

Figure 1: Tender purple nodule over the inner side of left forearm.

Figure 2: (A-C) Sections show dermal proliferation of perivascular small cells with round nuclei and eosinophilic cytoplasm separated by vascular channels; (D) Immunohistochemistry for MSA (muscle specific actin) is positive.

A 54 years old male, presented to the dermatology clinic with painful nodule over the left forearm for 15 years duration. The patient is known to have diabetes mellitus, hypertension, Ischemic heart disease, dyslipidemia, irritable bowel syndrome and positive history of colonic polyps. Examination of the skin revealed a single, tender and violaceous nodule measuring 1x1 cm over the anterior aspect of the left arm (Figure 1), with a minimal increase in size overtime. The lesion is not sensitive to cold. The patient denied the presence of similar lesion in other site of his body. In addition, there is no family history of similar illness.

The lesion was excised under local anesthesia and sent for pathology evaluation. Histopathology confirmed the diagnoses of glomus tumor (Figure 2).

Figure 3: Six months after excision.

Patient had followed up appointment after six months of excision and his symptoms resolved completely (Figure 3).

DISCUSSION

Glomus body is a specialized structure found in the reticular dermis and subcutaneous tissue, consisting of arteriovenous shunt that is made up of smooth muscle cells. Its function is to maintain adequate temperature and blood pressure by regulating blood flow. Glomus tumors are derived from the smooth muscle cells. Glomus bodies are predominantly found in the reticular dermis of the fingers, which reflects the most common site of these tumors in subungual area. A retrospective study of 138 cases revealed the distribution of glomus tumor, digits were involved in the majority of the cases with a percentage of 56. Arms and legs were the second sites affected with 14% and 13% respectively.

More interestingly, glomus tumor has been reported in extra-cutaneous locations that do not usually contain glomus body such as kidney, bone, nerve and mediastinum.

Glomus tumors are mostly benign with a few cases reported to be malignant, affecting males and females equally except the glomus tumor of the subungual area which favors middle aged women. These tumors are usually solitary especially when the fingers are affected.
Multiple tumors are often painless, making early and accurate diagnosis tremendously challenging.\textsuperscript{3}

Despite the classical symptoms of glomus tumor, its diagnosis is often delayed, with an average of seven years, mainly due to low index of suspicion and the variation of tumor location and presentations.\textsuperscript{1} Therefore, a suspicion of glomus tumor from history followed by specific physical examination is the core of diagnosis.

Clinically, patients present with painful, slightly raised, red to blue lesions with a size ranging from 0.1 to 3 cm in diameter.\textsuperscript{5} The diagnostic key of digital glomus tumor is the classic triad of severe pain, pinpoint tenderness and temperature hypersensitivity.\textsuperscript{4} However, extra digital glomus tumors do not always share the typical picture, specially for tumor size and the presence of temperature hypersensitivity.

Glomus tumor of the forearm often is larger and rarely sensitive to cold.\textsuperscript{1,2,5} These finding has been observed in other case reports of glomus tumor of the forearm and in our patient, who did not complain of cold intolerance and had a tumor size of 3 cm.\textsuperscript{6}

These symptoms can be examined by specific maneuvers, the Love test, Hildreth test and cold sensitivity test. Although, they have shown a very high sensitivity and specificity in diagnosing digital glomus tumor of the digits, there are no specific data of their accuracy in diagnosing glomus tumor of the forearm.\textsuperscript{6}

Radiography is not a diagnostic tool for glomus tumor.\textsuperscript{2} However, it can aid in the diagnosis. For example, ultrasound can help to differentiate between solid and cystic masses and localization of the tumor. MRI is useful to localize the tumor, determine its size and detect bone erosions but it less valuable in atypical locations and pathology.\textsuperscript{9} None of the imaging techniques was performed in our patient because glomus tumor was suspected based on the clinical presentation. Moreover, the only definitive diagnosis can be made by pathology evaluation, which confirmed the diagnosis of glomus tumor in our case.

Based on histopathology, glomus tumors contain three components, glomus cells, blood vessels and smooth muscle cells, and can be divided into three subgroups depending on the prominent structure. Solid tumors (few blood vessels and smooth muscles) glomangioma (high blood vessels component) and glomangiomyoma (high blood vessels and smooth muscles component). In both digital and extradigital lesions, Solid tumor is the most common type 75%, followed by glomangioma 20% and the least common is glomangiomyoma 5%.\textsuperscript{1} In our case, histopathology examination showed glomangioma variant, which is found to be significantly higher in extra-digital lesions than digital lesions.\textsuperscript{5}

Surgical excision with clear margins is curative and result in resolution of the symptoms.

In comparison to the recurrence rate of digital glomus tumor, which is reported to be between 12% to 33%, extra-digital lesions have a lower percentage with 10% recurrence rate.\textsuperscript{3} Early recurrence is suggested to be due to incomplete excision and late recurrence as result of development of a new tumor.\textsuperscript{4} Our patient underwent surgical excision which resolved the symptoms. Lastly, he was followed up one month later with no sign of recurrent symptoms such as pain or tenderness.

CONCLUSION

Glomus tumor are benign vascular tumor, but usually impact patients’ quality of life due to the severity of the pain and the persistence of symptoms for years before being diagnosed. Atypical locations and presentations of extra-digital tumors can lead to misdiagnosis, particularly where the lesion is rarely reported. Therefore, being aware of this variation and special characteristics of extra-digital tumors can lead to early diagnosis and treatment.

Finally, physical examination of painful subcutaneous mass in the limbs should always be conducted with the raise suspicion of glomus tumor.

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