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Chukwuemeka Ezeoke, Dong Xiang, Nishant Poddar

ABSTRACT

Introduction: This is the case of a patient with a symptomatic painful mass on right thigh. The patient was found to have a very rare case of glomus tumor of the thigh, which has been seen in only 2% of cases since this tumor was first discovered in 1924. A glomus tumor should be considered in the differential diagnosis of a painful, benign mass in a limb.

Case Report: A 48-year-old African-American morbidly obese female presented with a tender mass on the back of her right thigh. The mass progressively increased in size for three months prior to presentation. She denied having a mass under the nail bed, hands, palms, or any breast lumps. Her yearly mammograms were negative. Magnetic resonance imaging (MRI) scan showed a well-circumscribed mass measuring 3.1x2.2 cm. She was referred to a vascular surgeon for excisional biopsy, which revealed a glomus tumor of uncertain malignant potential measuring 2.7 cm extending to the inked surgical margin. She underwent re-excision with no evidence of residual mass.

Conclusion: Glomus tumor of the thigh is very rare. Pain, tenderness, and cold hypersensitivity should raise the suspicion for glomus tumor.

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CASE REPORT

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Introduction: This is the case of a patient with a symptomatic painful mass on right thigh. The patient was found to have a very rare case of glomus tumor of the thigh, which has been seen in only 2% of cases since this tumor was first discovered in 1924. A glomus tumor should be considered in the differential diagnosis of a painful, benign mass in a limb. **Case Report:** A 48-year-old African-American morbidly obese female presented with a tender mass on the back of her right thigh. The mass progressively increased in size for three months prior to presentation. She denied having a mass under the nail bed, hands, palms, or any breast lumps. Her yearly mammograms were negative. Magnetic resonance imaging (MRI) scan showed a well-circumscribed mass measuring 3.1x2.2 cm. She was referred to a vascular surgeon for excisional biopsy, which revealed a glomus tumor of uncertain malignant potential measuring 2.7 cm extending to the inked surgical margin. She underwent re-excision with no evidence of residual mass. **Conclusion:** Glomus tumor of the thigh is very rare. Pain, tenderness, and cold

hypersensitivity should raise the suspicion for glomus tumor.

Keywords: Glomus tumor of thigh, Glomus tumor with uncertain malignant potential, Glomus tumor

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INTRODUCTION

This is a case of a 48-year-old female who initially presented with a painful progressive enlarging mass on the back of her right thigh. The mass progressively increased in size with more discomfort at rest. She denied any pruritus, fever, erythema or other systemic symptoms. She was found to have a rare case of glomus tumor of the thigh, which has been seen in 2% of cases since this tumor was discovered in 1924. Often, missed at presentation, a glomus tumor should be considered in any painful, benign mass in a limb. The likelihood of the tumor progressing to malignancy is identified by a number of factors: size >2 cm, atypia, increased mitotic activity >2 per 50/hpf, high nuclear grade, and depth of involvement in relation to muscle fascia or visceral location [1].

CASE REPORT

A 48-year-old African-American female with a history significant for asthma, hypertension, and osteoarthritis, initially presented with a painful mass on the posterior right thigh. The mass progressively increased in size

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causing the patient to seek medical evaluation. Patient denied ever having a mass under the nail bed, hands, palms, or any breast lumps on yearly mammogram.

On presentation, her vital signs were normal. Physical examination was remarkable for an immobile, non-indurated, non-erythematous, 1x1.5 cm mass on the right posterior mid-thigh region with tenderness to palpation, and without any palpable lymph nodes. No abnormalities were seen on laboratory studies. Chest X-ray and electrocardiogram were without abnormalities.

Magnetic resonance imaging (MRI) scan of the right thigh which showed a well-circumscribed mass measuring 3.1x2.2 cm (Figures 1 and 2). She underwent resection of the painful mass with immediate relief of pain. Excisional biopsy grossly showed a 2.7x2.5x1.0 cm tumor surrounded by subcutaneous adipose tissue. It was well circumscribed and partially surrounded by a thin fibrotic capsule. Serial sectioning of the tumor showed a yellowish-tan, lobulated surface. There was no necrosis or hemorrhage identified grossly. The tumor appeared to be completely excised with grossly negative margins. Under low magnification, the tumor appeared to be well circumscribed and surrounded by a fibrous capsule (Figure 3). The lesion contained numerous irregular dilated vascular channels lined by bland single layer endothelial cells. Nests of neoplastic epithelioid cells surrounded the vascular channels. Tumor cells were present at the inked margin.

Under high magnification, epithelioid cells were mostly uniform in size with small round nuclei and inconspicuous nucleoli. The tumor cells contained moderate amount of eosinophilic cytoplasm. Mitotic rate was low (less than 2 per 50 high power fields). There was no cytological atypia or atypical mitosis identified (Figure 4). Immunohistochemical stains demonstrated that the tumor cells were positive for smooth muscle antibody (SMA), caldesmon, Desmin and CD34 and negative for S100 and AE1/AE3 (Figures 5–6). Ki-67 showed a mitotic index less than 1%. Five months later, she underwent re-excision as the inked margin of initial excisional biopsy was positive for tumor cells. She has been disease free with no clinical evidence of recurrence.

DISCUSSION

Glomus tumor is a well-circumscribed mass composed of vessels surrounded by epithelioid cells with uniform, round nuclei. It is very rare and comprises 2% of soft tissue tumors [2, 3]. The glomus body, a thermoregulator, is an arteriovenous anastomosis localized in dermal, precoccygeal soft tissue and in areas of the skin that are rich in glomus bodies (for example, the subungual regions of digits or the deep dermis of the palm, wrist, forearm, and foot) [2, 4].

Glomus tumor is a rare, benign neoplasm composed of cells resembling smooth muscle cells of the normal glomus body. Its clinical differential diagnosis includes:

Figure 1: MRI right femur: GRE Axial view of the glomus tumor.

Figure 2: MRI right femur: STIR Sagittal view of the glomus tumor.

Figure 3: A low power view showing that the tumor is well circumscribed with a fibrotic capsule.

Figure 4: A high power view showing the tumor contains numerous irregular vascular channels surrounded by nests of epithelioid cells with uniform nuclei, inconspicuous nucleoli, and eosinophilic cytoplasm.

Figure 5: Immunostain showing the tumor cells are diffusely positive for alpha smooth muscle actin.

Figure 6: Immunostain showing the tumor cells are positive for CD34.

abscess, epidermal inclusion cysts, hemangioma, arteriovenous malformations. Glomus tumor may be observed at any age. In most instances, it occurs in the fourth or fifth decades of life. 73% of glomus tumor occurs in the upper extremities; 23% in the lower extremities, of which only 2% occurs in the thigh [2, 4]. Presentation is classically a triad of pain, pinpoint tenderness, and hypersensitivity to cold. Localized pain and tenderness can be detected in 86% of these patients, but cold intolerance occurs in less than 2% [5, 6]. Vascular resistance through muscle contractions because of their intramuscular proximity decreases blood flow to the tumor. In essence, glomus tumors originate from neuro-myoarterial bodies; therefore, temperature changes will affect vascular resistance because of its thermoregulatory properties via skin blood flow, and cold simulations.

Ultrasound is useful in diagnosing cases in the outpatient clinic. MRI scan is used for accurate guidance and evaluation of tumor for excisional biopsy. Biopsy confirms the diagnosis. Histologically, glomus tumor is a well-circumscribed dermal nodule composed of glomus cells, vasculature, and smooth muscle cells. Solid glomus tumor, with scarce vasculature and scant muscle component, is the most common variant. Less common

variants include glomangioma, with prominent vascular component, and glomangiomyoma, with prominent vascular and smooth muscle components [1, 2, 3].

Glomus tumors are classified as: malignant with a 30–100% risk of metastasis, symplastic, glomangiomatosis, and tumor of uncertain malignant potential. Tumor of uncertain malignant potential is defined per College of American Pathologists as follows: superficial location and high mitotic activity or size >2 cm only, or deep location only. Our patient's tumor is classified as tumor of uncertain malignant potential because it is >2 cm, but superficial, with low mitotic activity [1, 2]. No case of glomus tumor of uncertain malignant potential has been reported to become metastatic or malignant after excision despite 7–10 years of follow-up; although one case did require re-excision [2]. Therefore, its benign nature is still uncertain. Close follow-up is advised. The most definitive treatment so far with the limited data available is surgical excision [2]. The risk of progression to malignancy is unknown, however, more cases need to be reported for better understanding of the disease and developing effective treatment to prevent recurrence.

CONCLUSION

Glomus tumor of the thigh is very rare. Pain, tenderness, and cold hypersensitivity should raise suspicion for glomus tumor.

Author Contributions

Chukwuemeka Ezeoke – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content; Final approval of the version to be published.

Dong Xiang – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Nishant Poddar – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published.

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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