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Malignant glomus tumor of the foot. Case report

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Glomangiosarcoma or is an extremely rare tumor. Few cases are published in the literature.**CASE PRESENTATION:** We present a rare case of 64-year-old female patient presented small reddish subcutaneous nodules on the sole of the foot. Surgical resection revealed malignant glomus, the evolution was marked by an infiltrating local recurrence leading to amputation, without notable metastases after six months.**CLINICAL DISCUSSION:** Malignant glomus tumor exhibit unusual characteristics, notably deep localization, large size and infiltration, mitotic activity, nuclear pleomorphism and mitonecrosis. The wide excision and possible amputation for infiltrating local type unresectable was the adequate treatment.**CONCLUSION:** Glomangiosarcoma arising de novo are exceedingly rare and must be considered the most aggressive and with high potential risk of metastasis.© 2021 The Author(s). Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

The majority of glomus tumors are small and benign. They develop from the neuromyovascular glomus located at the dermo-hypodermal junction. It is the main actor in skin microcirculation and thermal regulation [1]. These tumors are rare in the foot and are most often located in the hand, particularly the fingers. However, a malignant sarcomatous transformation of glomus tumors glomangiosarcoma can exceptionally occur [2]. The first reported case of an atypical and infiltrating aspect of a glomus tumor dates back to 1972 by Lumley and Stansfeld [3]. We report a case of an infiltrating malignant giant glomus tumor located on the sole of the foot.

This case report has been reported in line with the SCARE 2020 criteria [4].

2. Case report

A 64 years old woman followed for type II diabetes under well-balanced oral antidiabetics, without allergic or drug and no similar case in the family. She had for a few months presented small reddish subcutaneous nodules on the sole of her right foot (Fig. 1.a), she did not report any history of trauma, these nodules remained lit-

tle symptomatic until she consulted in a private hospital structure where she underwent a biopsy-resection of the tumor. Six months later, the patient consulted in our hospital for a recurrence of the tumor on the sole of the foot, the physical examination noted a multilobed red-bluish mass of about 8 cm, with a few of nodule tumor, very painful at the slightest pressure, with a dorsal extension through the inter-metatarsal space (Fig. 1.bc), she could not support the footwear and any direct contact, walking was impossible, the symptoms were not alleviated by medical treatment, no regional lymphadenopathy was noted, the standard X-ray showed an increase in the density of the soft tissues without bone abnormalities, the MRI showed a polylobed tumor process centered on the plantar surface of the foot with dorsal extension (Fig. 2), with a heterogeneous and compartmentalized signal, locally advanced, site of areas with T1 hypersignal and T2 hypersignal fluid, and areas with T1 and T2 hypointense slightly enhanced after injection of gadolinium that does not disappear after removal of fat, with an extension of the first metatarsal to 5th metatarsal with the presence of two rounded processes with the same characteristics at the level of the cuboid and medial cuneiform. Histological examination of the first surgical resection of the four nodules revealed nodular tumor proliferation, it forms large nodules, incurred by thick fibrous tracts. Some nodules are carved out of large vascular caverns, cavernous, filled with red blood cells. Tumor cells are round with rounded nuclei with numerous mitotic figures (32 mitoses / 50fields), as well as some mitonecrosis (Fig. 3).

The immunohistochemical study showed that tumor cells most often expressed smooth muscle actin, and focal caldesmosis and

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Fig. 1. (A): Clinical appearance of the foot tumor. (B, C): Appearance after tumor recurrence.



Fig. 2. Polylobed infiltrating aspect of the foot tumor on MRI.

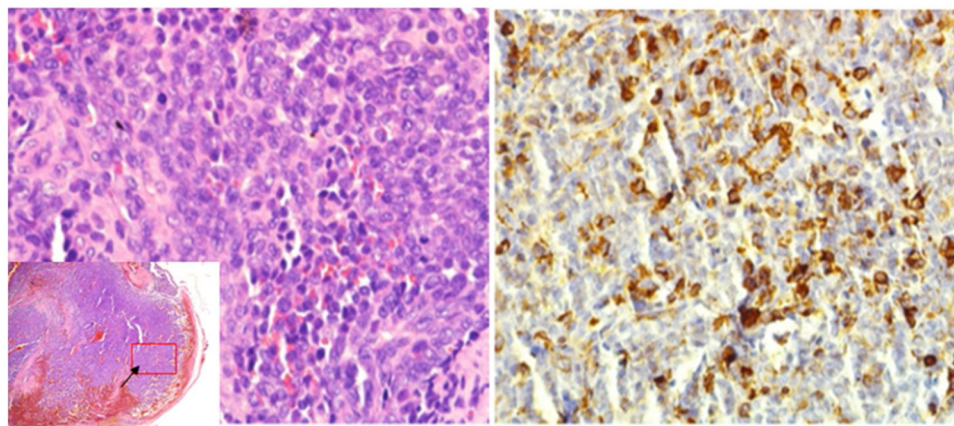


Fig. 3. Tumor proliferation showing numerous mitoses, and positivity of tumor cells to anti-smooth muscle antibody (Immunohistochemistry, ×40).

synaptophysin, they did not express CD34, P-S100 and HMB-45. The appearance is more in favor glomangiosarcoma arising de Novo. The extension workup was negative (no metastases). Due to the aggressiveness of the tumor and the advanced infiltration in width and depth, a carcinological resection proves to be impossible especially since the tumor is chemo and radio-resistant, a mid-leg amputation was performed in our center specializing in orthopedic and tumoral surgery by experienced operator. The evolution was simple without recurrence or metastases after a follow-up of 6 months with complete resumption of activities using leg prosthesis.

3. Discussion

The first description of a glomus tumor was made in 1872 by Wood [5] as a small painful subcutaneous nodule, of a benign nature. Masson [6] linked it in 1924 to a glomic origin. Glomus tumors originate from the glomus, a distinct physiologic arteriovenous anastomosis in the dermohypodermal junction involved in thermoregulation. The sarcomatous transformation is exceptional,

Lumley and Stansfeld [3] reported the first description of a malignant glomus tumor in the leg in a 24-year-old woman, one year later, Anagnostou et al. [7] reported a case of an infiltrating subcutaneous glomus tumor of the hand which he claimed to be a malignant tumor. In 1988, Aiba et al. [8] described a case of malignant glomus tumor characterized by pleomorphic cells with high mitotic activity located in the back in a 65-year-old man, the evolution was good after surgical resection. These malignant glomus tumors have been well elucidated by the work of Gould et al. [9] in 1990; they analyzed six cases of atypical glomus tumors and proposed a classification with three categories:

- Locally infiltrative glomus tumor “LIGT”: they are histologically identical to benign glomus tumors but with infiltrating edges.
- Glomangiosarcoma arising in a benign typical glomus tumor “ABC”: sarcomatous tumors originating from pre-existing benign glomus tumors (transformations)
- Glomangiosarcoma arising de novo “GADN”: the histological characteristics show a cellular pleomorphism with high mitotic

activity and local aggressiveness. This is the type our patient presents.

In 2001, Folpe et al. [2] analyzed a series of 52 atypical glomus tumors and they proposed criteria to classify them into 4 groups, Malignant glomus tumors: tumor with a deep localization and a size greater than 2 cm, or an atypical mitotic figure, or high mitotic activity > 5 mitoses / 50 fields. Symplastic glomus tumors: A high nuclear grade tumor in the absence of other malignant features. Glomus tumors with uncertain malignant potential: tumor devoid of the above criteria but having high mitotic activity and only superficial localization, or only large size, or only deep localization. Glomangiomas: tumor with histological features of diffuse angiomas and excess glomus cells. Our patient presented all criteria for malignancy, deep localization and giant size, high mitotic activity and cellular atypia.

Immunohistochemically, glomangiosarcomas are similar to benign glomus tumors, they express smooth muscle actin, vimentin, caldesmon [10]. Other markers including desmin, CD34, cytokeratin and S100 protein are usually negative as in our case. In 2005, T. Pérez de la Fuente [11] reviewed 25 cases reported as malignant glomus tumors since its first description by Lumley, 68% of cases were over 45 years old. Localization in the lower limb is the most common, other locations are also found including the upper limb, trunk and face. We founded only four cases of the hand and only one case of the foot [2,9,11–13]. There was no predominance of sex (sex ratio = 1). Benign glomus tumors on the other hand are the prerogative of the hand, the location in the lower limb remains exceptional, with a predominance of women, we have published in our department 2 unusual locations in the knee of benign glomus tumors revealed by pain of the knee not calmed by medical treatment [14]. Malignant glomus tumors tend to recur locally, and the risk of metastases is always present, five cases published in literature [15–19], all occurring in de Novo glomangiosarcoma. The first case of metastasis was reported by Kreutz [15] at the maxillary level from a large primary (10 × 5 × 5 cm) in the thigh, in Folpe's series, the metastasis rate was 38% (8 cases among 21 cases of malignant glomus tumors) [2].

Wide local excision remains the treatment of choice for this tumor in literature. In our case, the patient accepted our surgical procedure in front of the deep localization and the impossibility of carcinological resection. We insist on the essential value of systematic biopsy in front of any mass before surgical resection, in order to establish an early diagnosis and adequate therapeutic management with histopathologically tumor-free margins, due to the risk of malignant transformation of glomus tumors even if this is exceptional but remains possible. Radiotherapy or chemotherapy has not found its place in the few cases published, except for one reported by Milia et al. [20] of malignant glomus tumor in a 40 year-old man presenting a lesion in the upper cervical region, having combined radiotherapy and chemotherapy with successful outcome.

4. Conclusion

Malignant glomus tumor is exceedingly rare and the localization on the foot was exceptional, the diagnosis is based on the histopathological examination. The wide excision and possible amputation for infiltrating local type unresectable was the adequate treatment. The group of glomangiosarcomas arising de novo considered the most aggressive of these tumors with high potential risk of metastasis.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

This research did not require ethical approval due to the institute not requiring it for this type of study.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying image.

Author contribution

Zakaria Rkiba: Corresponding author writing paper.
Mohamed Rafai: supervised the writing of the manuscript.
Abdessamad Rajaallah: study concept.
Charafeddine Elkassimi: study concept.
Abdelhak Garch: correction of the paper.

Registration of research studies

Not applicable.

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DR Zakaria Rkiba.

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