

## **Giant Malignant Schwannoma in the Right Thoracic Limb: Case Report**

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### **ABSTRACT**

**Introduction:** Tumors of the sheath of the peripheral nerves, also known as schwannomas, mainly benign. The malignancy of these is very rare and there are few cases reported in the literature.

**Clinical case:** A 62-year-old male with a diagnosis of ulcerated malignant schwannoma in the distal third of the posterior aspect of the right thoracic limb who attended the oncosurgery service. The CT scan reported a large solid tumor, predominantly hypodense, with a probable origin dependent on the poorly defined triceps brachii muscle. Surgical removal was performed with two centimeters of perilesional margins, performing primary closure with drain placement.

**Conclusion:** Surgical treatment of total surgical removal with adequate margin coverage is the treatment of choice as a protective factor in survival in this type of tumor.

### **ARTICLE DETAILS**

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### **INTRODUCTION**

Schwannomas, or also known as neurilenomas, are benign tumors derived from the myelin sheaths of peripheral nerves and represent approximately 5% of soft tissue tumors of the upper limb.<sup>1,4</sup> They occur spontaneously and are mainly associated with family history, with neurofibromatosis type 2, schwannomatosis and Carney complex.<sup>3</sup> According to the World Health Organization, they are grade I tumors and very rarely become malignant, mainly in malignant tumors of the peripheral nerve sheath or angiosarcoma.

### **CLINICAL CASE**

A 62-year-old male, pensioner, with a significant history of schizophrenia diagnosed 10 years ago on treatment with clozapine, benign prostatic hyperplasia for 3 years on treatment with tamsulosin, hypercholesterolemia for 3 years on treatment with pravastatin. Positive smoking at a rate of 1 pack a day for 50 years, stopped 1 month ago, with a smoking index of 50. He denies any history of surgery or allergies.

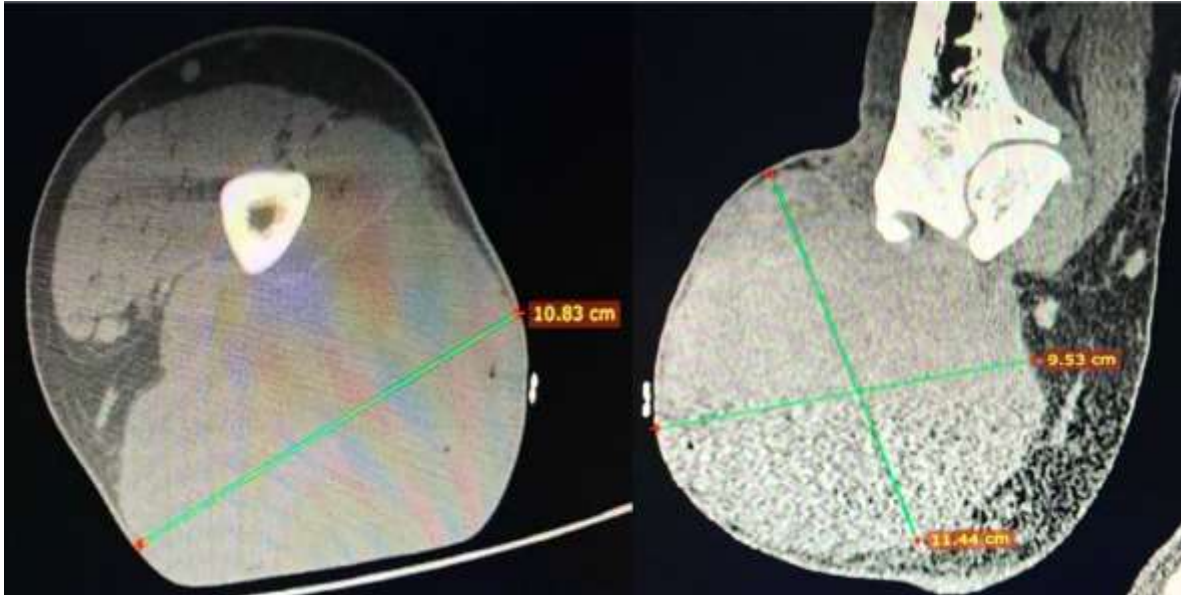


**Image 1. Ulcerated solid tumor on the posterior aspect of the right thoracic limb.**

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He refers to starting 8 months ago with the presence of a tumor less than 1 cm in the posterior region of the right elbow, painless, with a subsequent increase in size and involuntary weight loss of approximately 30 kg in 8 months, which is why he went for evaluation by oncological surgery where On physical examination, a solid tumor was identified in the region behind the right elbow, fixed, painless, with irregular edges of approximately 15 cm by 15 cm, with a non-bleeding ulcer in the center. No axillary, cervical, or inguinal lymph

nodes are palpable. A CT scan of the right thoracic limb was requested, reporting a solid tumor of 11.4 x 9.5 cm dependent on the triceps muscle with a poorly defined hypodense area, accompanied by acute inflammation of all the fatty tissue and thickening of the skin (Images 2-3). Closed biopsy with Trucut 16G was performed with histopathological report of high-grade malignant spindle cell neoplasm. Therefore, wide resection is electively decided. Normal preoperative laboratories



**Images 2-3.** CT scan of the right thoracic limb reporting a solid tumor measuring 11.4 x 9.5 cm dependent on the triceps muscle with a poorly defined hypodense area, accompanied by acute inflammation of all the fatty tissue and thickening of the skin.

### Surgical technique

Preoperative marking was performed with 2 cm margins (image 1), then a spindle incision was made with a 3:1 ratio of the lesion, it was approached by planes, dissecting with wide margins, predominantly between the triceps brachii muscle fibers, performing complete resection of the lesion

(images 4-5). Widely, 2 cm perilesional undermining was performed, a ¼-inch Drenovac type drain was placed, subcutaneous and deep tissue was faced with simple separated sutures with 2-0 vicryl and skin was faced with 2-0 nylon with Sarnoff stitches.



**Images 4 and 5.** Resected tumor with 2-cm margins and its relationship with the surgical bed. Integrity of the ulnar nerve can be observed.

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During the postoperative period, blood flow was approximately 50 cc per day. Discharged with adequate improvement two days after the operation, withdrawing the DRENOVAC at 7 days with little serous output. (Image 6). Currently under follow-up by Medical Oncology for adjuvant treatment with radiotherapy. The histopathological report described a lesion with a white nodular appearance, firm in

consistency, neoplastic in appearance, measuring 12.3 cm by 9.5 cm, with a capsule of loose fibroconnective tissue, lobulated in appearance with central areas of fibrous appearance, diagnosing grade 2 ulcerated malignant Schwannoma of the French classification of sarcomas, free surgical margins and no evidence of lymphovascular infiltration.



**Image 6. Postoperative wound closure with Sarnoff type suture and placement of DRENOVAC.**

### DISCUSSION

Malignant schwannoma, also known as neurofibrosarcoma, neurogenic sarcoma, malignant neurilemoma, or malignant peripheral nerve sheath tumors, is a rare entity, with an approximate incidence of 0.001% in the population.<sup>8</sup> Although schwannomas are the most common type of benign nerve sheath tumors. Only a very small number of peripheral nerve sheath tumors have been previously reported as schwannomas.<sup>10</sup> This phenomenon was first reported in 1994 by Woodruff et al.,<sup>11</sup> who reported 2 cases of peripheral malignancy of nerve sheath tumors. The malignant lineage is a rare malignant mesenchymal lesion with a prevalence of 5% of all soft tissue sarcomas.<sup>6</sup> These are mainly related to autosomal dominant genetic syndromes such as neurofibromatosis types 1 and 2.<sup>3,5</sup> It has been described that patients with neurofibromatosis type 1 have greater risk of malignant transformation in pre-existing plexiform neurofibroma.<sup>8</sup> In general, they are known to have high metastatic potential and poor prognosis. Multiple series of studies report a 5-year survival of 15-50%.<sup>9</sup> The most common sites involve nerve roots and bundles in the

extremities and pelvis, mainly the sciatic nerve. Up to 50% of patients have metastatic disease, mainly to the lung.<sup>9</sup> Cai et al. reported that the location with the highest risk of poor prognosis is in the head and neck compared to the location in the extremities.<sup>6</sup> Most schwannomas are asymptomatic, such as pain, weakness and paresthesias. Due to its presentation in the upper extremities, it is common to initially think of other differential diagnoses such as lipomas, synovial cysts, ganglions and other pathologies.<sup>2</sup> Magnetic resonance imaging is the most useful imaging study to determine the anatomic extent and the surgical plan. In the setting of localized disease of soft tissue sarcomas, complete surgical excision with clear margins is the treatment of choice. Tumor size (considered larger than 5 cm) is considered the most determining prognostic factor in the literature. Other factors reported are tumor grade, truncal location, surgical margins, local recurrence, and heterologous rhabdomyoblastic differentiation.<sup>9</sup> Free margins are of vital importance for the control of local recurrence, as well as survival.<sup>4</sup> Therefore, according to the literature, free margins are a protective factor for a better prognosis.<sup>6</sup>

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### CONCLUSION

The surgical treatment of choice for peripheral nerve sheath tumors is surgical removal with free margins, since this determines a better prognosis in patient survival.

### CONFLICT OF INTERESTS

The authors declare that they have no conflict of interest.

### Ethical Responsibilities

Protection of people and animals. The authors declare that no experiments were carried out on humans or animals for this research.

Data confidentiality. The authors declare that they have followed the protocols of their work center regarding the publication of patient data.

Right to privacy and informed consent. The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

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