RADIATION-INDUCED SARCOMAS: A SURGICAL CHALLENGE

SARCOMAS INDUZIDOS POR RADIAÇÕES: UM DESAFIO CIRÚRGICO

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ABSTRACT

Malignant peripheral nerve sheath tumors (MPNST) are uncommon, biologically challenging soft tissue sarcomas. The definitive treatment of these tumors is extremely challenging. In 50% of cases, these tumors are associated with type I neurofibromatosis, but in some cases, the cause can be previous treatment with radiotherapy. The present article pretends to report a clinical case of a radiation-induced MPNST submitted to surgical treatment.

***Keywords**: MPNST, radiotherapy treatment, radiation-induced sarcomas, multimodality treatment.*

RESUMO

Os tumores das bainhas nervosas periféricos malignos (MPNST) são tumores raros e biologicamente agressivos, sendo o seu tratamento radical um desafio. Ocorrem em 50% dos casos em contexto de neurofibromatose tipo I. Uma outra causa para estes tumores é a exposição a radioterapia prévia. O presente artigo relata um caso clínico de um MPNST radio-induzido submetido a tratamento cirúrgico.

***Palavras-chave:** MPNST, radioterapia, sarcomas induzidos por radiações, tratamento multimodal.*

BACKGROUND

Malignant peripheral nerve sheath tumors (MPNST) are rare single cell malignancies that account for 5 to 10% of all soft tissue sarcomas.[1]

The main risk factors for the development of these lesions are preexisting benign plexiform neurofibromas, prior radiation therapy and large germline mutations involving the entire NF1 gene.[2,3]

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Adjuvant radiotherapy has an extremely important role in the treatment of numerous neoplasms, such as breast cancer. This treatment reduces local recurrences, however, exposure to radiotherapy can induce, in rare cases, the formation of soft tissue sarcomas. Radiation-induced sarcomas are rare and they have a poor prognosis with an important risk of local relapse.[4,5]

Complete surgical excision with negative margins remains the only proven curative treatment.[5]

There is limited data about the advantages of using neoadjuvant treatment in this type of tumors but some patients might benefit with this treatment for downsizing and downstaging. Neoadjuvant radiotherapy is recommended for high-grade lesions or tumors greater than 5 cm. Prospective studies are needed in order to establish the potential benefit of neoadjuvant chemotherapy.[5]

The present article pretends to report a clinical case of a radiation-induced soft tissue sarcoma.

CLINICAL CASE

The authors present a case of a 51 year-old woman, with previous history of breast cancer treated in 2007 with surgery (partial mastectomy and axillary dissection) followed by adjuvant treatment with radiotherapy, chemotherapy and hormonal therapy. Consequently, after surgery the patient developed lymphedema and functional limitation of the arm. In November 2020, the patient complained about an infraclavicular mass with an irradiated pain to the arm, maintaining the lymphedema. The patient was initially studied with a CT scan that revealed a 6 cm lesion in contact with vascular-nervous bundle in the middle of the clavicle (Fig. 1).

For better characterization of the lesion, the patient was then submitted to a MRI that revealed an oval lesion with 70 x 40 x 43 cm near to brachial plexus, posteriorly to pectoral minor, with no evidence of arterial ou venous invasion (Fig 2). Even though there were no imaging evidence of vascular-nervous bundle invasion, the clinical evaluation was suggestive of brachial plexus involvement.

The biopsy was compatible with a malignant peripheral nerve sheath tumor (MPNST).

The case was discussed in the multidisciplinary sarcoma tumor board and the patient began neoadjuvant radiotherapy.

Neoadjuvant treatment was finished in April 2021 and the patient was re-evaluated with an MRI. The

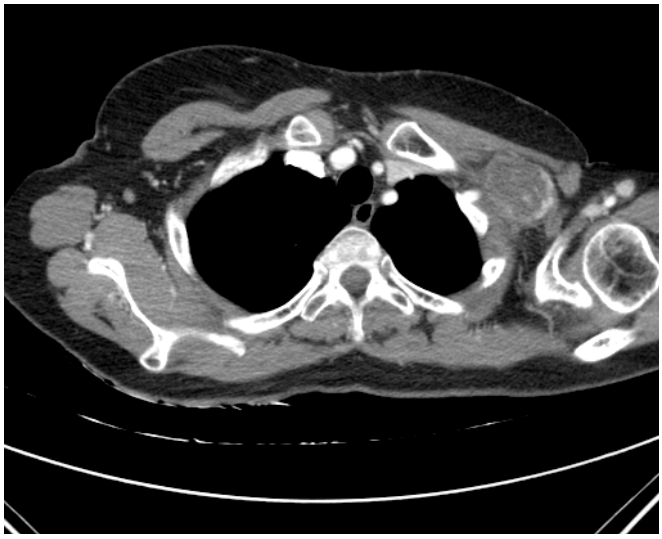

Fig. 1 – **CT Scan.**

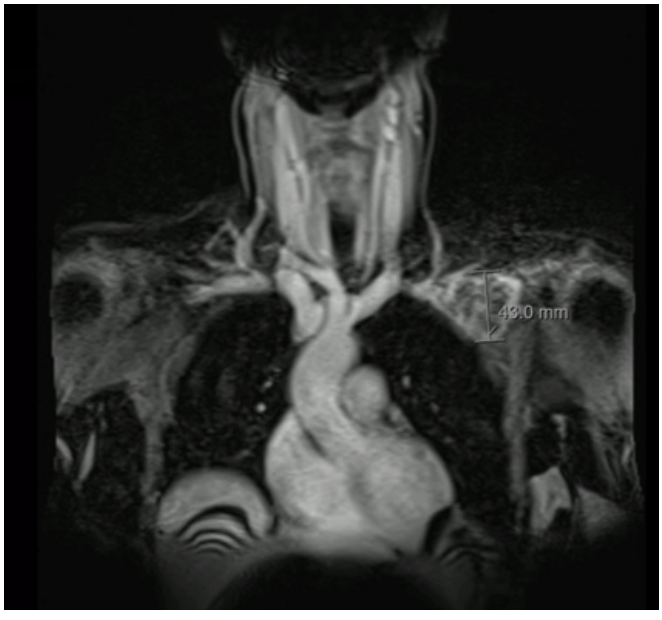

Fig. 2 – **Diagnostic MRI.**

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MRI revealed a lesion with lobular limits posterior to pectoralis major, with pectoralis major and minor invasion with contrast caption. This lesion had 75 x 42 x 47 mm and extends medially posterior to the clavicle. This exam revealed suspicion of subclavian vein invasion with intraluminal component in a 30 mm extension. The internal jugular vein, the left brachiocephalic vein and the superior segments of cava had normal permeability. In the posterior surface of the mass, it contacted the brachial plexus. In the superior surface, the lesion was in contact with the clavicle but with no evidence of bone invasion. There were no axillary adenopathies (Fig. 3).

To complete the treatment the patient was submitted to surgical excision in May 2021.

To access the tumor, it was necessary to disarticulate sternoclavicular articulation. Then, left brachiocephalic, left subclavian and left internal jugular veins were identified (Fig. 4).

There was an evident muscular invasion of major and minor pectoralis and invasion of subclavian vein near its origin in left brachiocephalic vein. There was no evidence of arterial invasion. This neoplasm seemed to have its origin in nervous sheath of the brachial plexus.

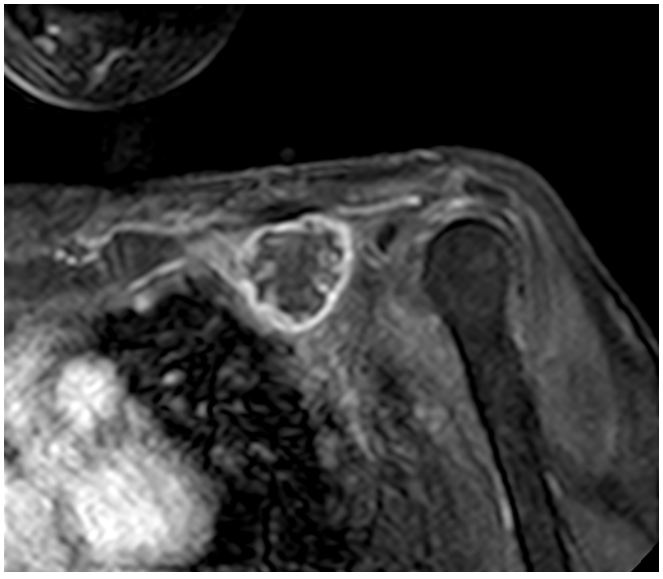

Fig. 3 – **Re-evaluation MRI after neoadjuvant radiotherapy.**

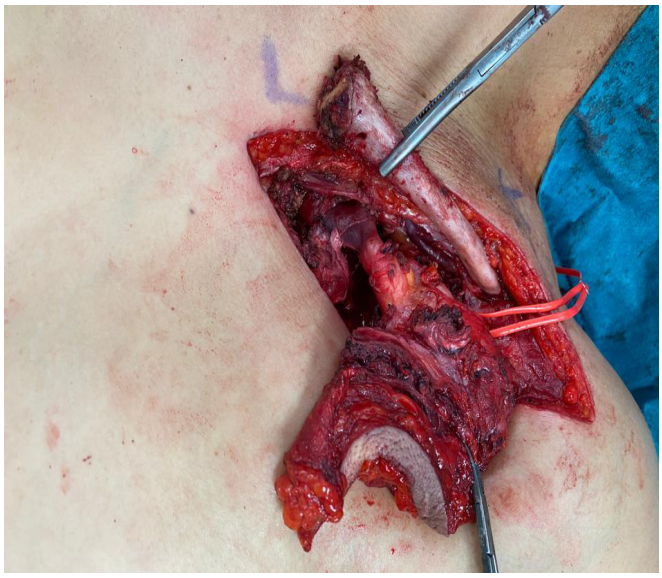

Fig. 4 – **Exposure of the tumor with involvement of the subclavian vein after disarticulation of clavicle.**

The tumor was dissected with preservation of subclavian artery and brachial plexus, with section of the fiber that seemed the origin of the tumor. The thoracic duct was ligated and subclavian vein was sectioned proximally in its origin near the left brachiocephalic vein and distally to the tumor allowing tumor resection (Fig 5).

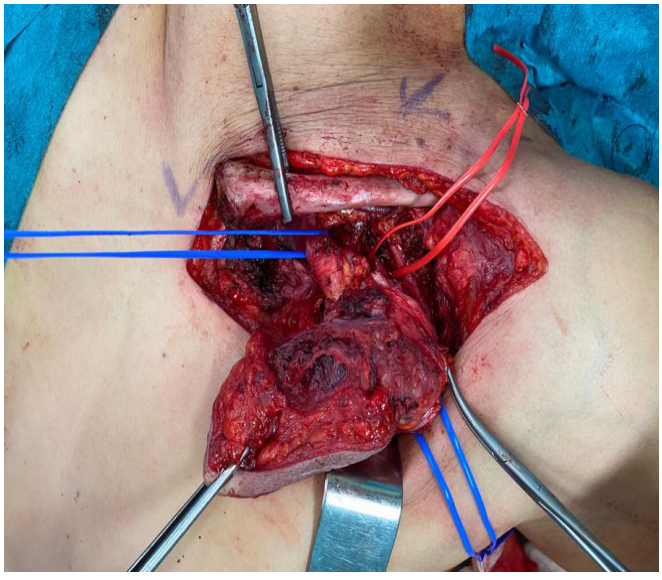

Fig. 5 – **Dissection and identification of left subclavian vein and artery proximally and distally to the tumor.**

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The clavicle was then fixated and, to protect brachial plexus and subclavian artery, some fibers of pectoralis major were transposed superiorly. An aspiration drain was placed (Fig. 6).

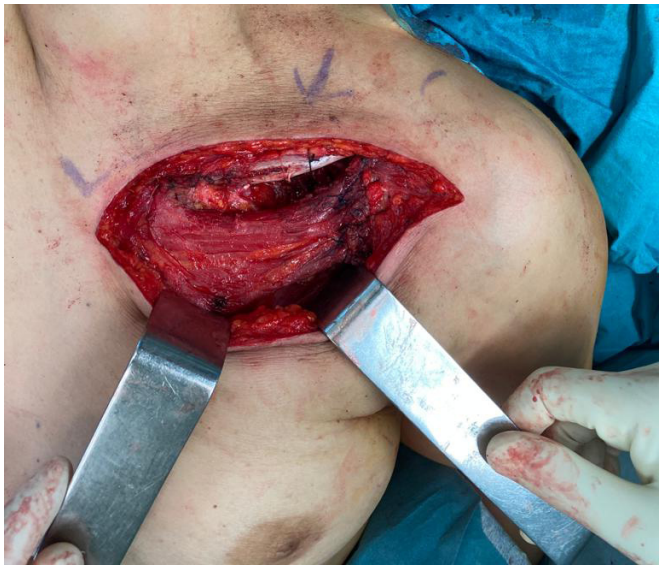

FIG. 6 – **Fixation of the clavicle and muscle transposition.**

Postoperative period had no adverse events. The patient was evaluated by a physiatrist to start rehabilitation treatments and was discharged from the hospital six days after the procedure.

Surgical specimen confirmed a MPNST compatible with previous biopsy, with osteocartilaginous and leiomyossarcomatous differentiation secondary to radiotherapy treatment.

The case was evaluated in a multidisciplinary sarcoma tumor board and it was decided for clinical and imagiological surveillance.

The patient is in follow-up for 18 months with no evidence of tumor relapse. The patient maintains upper limb lymphedema and functional limitation improving with rehabilitation treatment.

FINAL CONSIDERATIONS

Radiation-induced MPNST have a worse prognosis than sporadic lesions.

Despite the fact that neoadjuvant treatment in this setting remains in debate, patients with high grade tumors, tumors greater than 5 cm and with invasion of important structures might benefit with neoadjuvant radiotherapy.

Surgical treatment remains the curative treatment for these soft-tissue sarcomas.

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