www.journalofcancerology.com PERMANYER J Cancerol. 2015;2:113-6

JOURNAL OF CANCEROLOGY CASE REPORT

Intimal Sarcoma of the Basilic Vein and Hodgkin's Lymphoma

JORGE LUIS MARTÍNEZ-TLAHUEL[1*], LILIANA IVETT HERNÁNDEZ-CRUZ[2] AND JAIME DE LA GARZA-SALAZAR[1]

[1]Instituto Nacional de Cancerología (INCan), Mexico City, Mexico; [2]Hospital Regional Rafael Pascacio Gamboa, Tuxtla Gutiérrez, Chiapas, Mexico

ABSTRACT

Soft tissue sarcomas are a group of rare solid tumors with distinct clinical and pathological features. They arise from mesenchymal tissue with heterogeneous differentiation. They account for less than 1% of all malignant tumors. Sarcomas of the great vessels usually present in the aorta, pulmonary artery, and inferior vena cava. Peripheral arterial sarcomas are exceptionally rare. They have been reported in the iliac and common or deep femoral arteries and are frequently undifferentiated. The majority of peripheral arterial sarcomas are leiomyosarcomas or angiosarcomas. Primary neoplasms of the major blood vessels are divided into three categories based on their site of origin: large veins, pulmonary artery, aorta artery, and its branches. In the following report we present the case of a male in his thirties with a history of Hodgkin's lymphoma, treated with first- and second-line chemotherapy regimens, who achieved a complete response after treatment, with no evidence of disease. Then in 2014 he was diagnosed with intimal sarcoma of the basilic vein in the left arm and treated with surgery as first line of treatment followed by adjuvant chemotherapy and radiation therapy. He later presented with lung metastases so metastasectomy was performed. The patient is currently being followed. The purpose of this paper is to present the case as there are not many reports on cases of this type and the site of presentation is unusual. The diagnostic approach is also described. Patients who receive radiation therapy as part of treatment for lymphoma are predisposed to develop sarcomas, which was not the case in this patient, so that this association may be explained by the presence of a genetic syndrome. (J CANCEROL. 2015;2:113-6)

Corresponding author: Jorge Luis Martínez-Tlahuel, dr.jorgetlahuel@gmail.com

Key words: Rare neoplasm. Intimal sarcoma. Basilic vein. Hodgkin's lymphoma.

Correspondence to:
*Jorge Luis Martínez-Tlahuel
Servicio de Piel y Partes Blandas
Instituto Nacional de Cancerología
México, D.F.
E-mail: dr.jorgetlahuel@gmail.com

Received for publication: 23-05-2015
Accepted for publication: 30-05-2015

INTRODUCTION

Soft tissue sarcomas are a group of rare tumors that arise from mesenchymal tissue with heterogeneous differentiation. They account for less than 1% of all malignant tumors[1-3]. Worldwide, the incidence rate is between 1.8 and 5.0 cases per 100,000 people per year[1]. The estimated number of new cases and deaths from soft tissue sarcoma in the United States in 2015 is reported to be 11,930 and 4,870, respectively[4].

Most are sporadic; few have an identifiable cause[3]. The truth is that little is known about the etiology of soft tissue sarcomas. Risk factors such as radiation and phenoxy herbicides or chlorophenols have been identified in only a small fraction of all cases. However, these findings are not supported by other studies. Genetic syndromes are a cause for cancer due to various mutations in different genes; some of these, such as Li–Fraumeni syndrome[1,3,5], Werner syndrome, and possible unidentified syndromes have been associated with a hereditary predisposition for the development of soft tissue sarcomas in a small percentage of all cases of soft tissue sarcoma[1]. Sarcomas may develop 3-5 years after treatment with radiation therapy for lymphoma, cervical cancer, or testicular tumors[3].

Sarcomas of the great vessels usually present in the aorta, pulmonary artery, and inferior vena cava. Over 60% of sarcomas of the aorta and its branches are intimal sarcomas, arising from the tunica intima of the large vessels[6,7]. Peripheral arterial sarcomas are exceptionally rare. They have been reported in the iliac and common or deep femoral arteries and are frequently undifferentiated. The majority of peripheral arterial sarcomas are leiomyosarcomas or angiosarcomas[6].

Here, we describe a case of a patient with an intimal sarcoma of the basilic vein, which is a rare neoplasm, and its management.

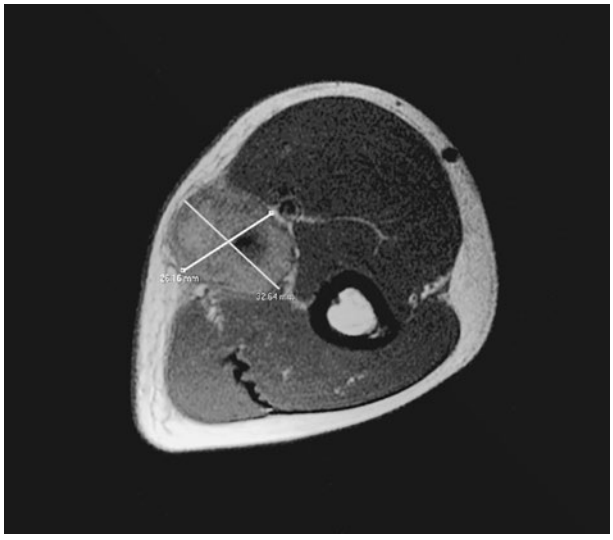

Figure 1. Magnetic resonance imaging. Left arm. Lesion of the medial compartment in the middle third of the left arm apparently attached to the basilic vein, with the actual tumor measuring approximately 5 × 3 cm along its different axes.

CASE REPORT

A 30-year old man who in 2004 presented with mixed-cellularity classical Hodgkin's lymphoma, clinical stage IIIB, received six cycles of ABVD (Adriamycin, bleomycin, vinblastine, dacarbazine). After being disease-free for one year and six months, he relapsed in March 2007. Second-line treatment with alternating cycles of ABVD and MOPP (mechlorethamine, vincristine, procarbazine, and prednisone) was initiated. A positron emission tomography-computed tomography (PET-CT) scan at the end of treatment in 2008 showed no tumor activity. In February 2014 the presence of a deep, indurated tumor, measuring approximately 4 × 3 cm was noted in the medial aspect of the left arm. A magnetic resonance imaging (MRI) was performed on his left arm, which showed a lesion of the medial compartment in the middle third of the left arm attached to the basilic vein, with the actual tumor measuring approximately 5 × 3 cm along its different axes (Fig. 1). Ultrasound-guided biopsy of the lesion was performed. The histopathological report revealed a high-grade sarcoma with pleomorphic areas and production of osteoid matrix

consistent with intimal sarcoma and negative for CD34. Wide surgical resection was the first-line treatment of the cancer, followed by chemotherapy with doxorubicin and ifosfamide for six cycles and adjuvant radiotherapy to the surgical bed (60 Gy in 30 fractions). Despite treatment, locoregional spread of the disease as well as lung metastases were detected in January 2015 so treatment with sunitinib was initiated. Metastasectomy was performed. The surgical approach was a right posterolateral thoracotomy and a wedge resection. The histopathological report revealed metastatic high-grade pleomorphic sarcoma.

At present the patient continues to be monitored and followed up.

DISCUSSION

Undifferentiated luminal sarcomas that could not be classified histologically as leiomyosarcoma, angiosarcoma, malignant fibrous histiocytoma, or osteosarcoma were called "intimal", although storiform growth and osteosarcoma were seen focally. An undifferentiated sarcoma involving the vessel wall was classified as pleomorphic[7].

Angiosarcomas that originate in the endothelium and mural tumors derived from the media and adventitia represent the majority of peripheral arterial sarcomas. Histology and immunohistochemistry are used to differentiate intimal sarcomas from these other two. Microscopy shows the endothelial cells in angiosarcomas and they are positive for vimentin (mesenchymal differentiation marker), an antigen related to factor VIII and CD34 (vascular markers). Leiomyosarcomas are characterized by spindle-shaped cells and are actin and desmin positive (muscle markers)[8]. Intimal sarcomas are of unknown cell origin and usually exhibit undifferentiated cells with variable immunophenotypic features[6]. Burke, et al. conducted a retrospective clinicopathologic study of 43 cases of sarcomas of the great vessels: 11 sarcomas of the aorta, 16 sarcomas of the inferior vena cava, and 16 sarcomas of the pulmonary artery, in which the sarcomas were negative for desmin, factor VIII-related antigen, S-100 protein, and CD34[7].

The clinical presentation of these tumors is nonspecific. The most common finding at presentation is a painless mass with gradual growth[3]. Sarcomas of the great vessels tend to occur early in patients with a history of peripheral vascular disease and mostly present at advanced stage and with an aggressive course[6].

Clinical examination, imaging, and histological analysis are required for the identification of soft tissue sarcomas. Physical examination and imaging can be used to define the relationship of the tumor to other structures.

Computed tomography and magnetic resonance imaging are used; MRI provides the best possible anatomic definition and is preferred for the diagnosis of sarcomas in the soft tissue of limbs. Percutaneous core needle biopsy is safe and effective. The subtype and grade of the tumor can be determined in 80% of biopsies with this procedure[3].

The treatment of choice consists of surgical resection with wide margins, with or without radiation therapy. Whereas the aim of surgery and radiation therapy is local tumor control, the aim of chemotherapy is systemic control, which may be therapeutic, adjuvant, or palliative, with doxorubicin and ifosfamide-based regimens. Although some subtypes of soft tissue sarcomas are sensitive to chemotherapy, the outcomes of therapeutic chemotherapy are unsatisfactory and the use of adjuvant chemotherapy is controversial. The role of chemotherapy and radiotherapy after surgical resection has not yet been proven[9]. A meta-analysis of adjuvant chemotherapy showed an improvement in progression-free survival, but did not show an overall survival advantage[3].

In the case of sarcoma of the pulmonary artery, the effectiveness of radiation therapy and chemotherapy

remains controversial, although some studies suggest that both therapies can be effective. Using these therapies after surgical treatment prolongs the survival time compared with surgery alone[10]. Large series confirm that grade and size have similar prognostic significance[3]. Despite advances in surgery, chemotherapy, and radiotherapy, the survival rate at one year for sarcomas of the great vessels has remained at 13%, with a mean survival of 14 months[6].

CONCLUSION

This case in particular describes a rare site for a sarcoma. We cannot associate the history of lymphoma with its etiology as the patient did not receive treatment with radiotherapy. The relationship between the two neoplasms may be explained by the presence of a genetic disorder that predisposes to the aforementioned syndromes, characterized by the development of multiple neoplasms.

In this case and as mentioned in the literature, the definitive diagnosis was established through histopathology, which showed pleomorphic areas and production of osteoid matrix, and CD34 immunohistochemistry, which was used mainly to differentiate it from other types of sarcoma such as angiosarcoma. Adjuvant chemotherapy is not considered the standard treatment for sarcomas, the basic regimen consisting of anthracyclines in combination with ifosfamide. Generally speaking, the prognosis for these patients is poor.

REFERENCES

1. Wibmer C, Leithner A, Zielonke N, Sperl M, Windhager R. Increasing incidence rates of soft tissue sarcoma? A population-based epidemiologic study and literature review. Ann Oncol. 2010;21:1106-11.
2. Sinha S, Peach AH. Diagnosis and management of soft tissue sarcoma. BMJ. 2010;341:c7170.
3. Clark MA, Fisher C, Judson I, Thomas JM. Soft-Tissue sarcomas in adults. N Engl J Med. 2005;353:701-11.
4. American Cancer Society: Cancer facts and figures 2015. Atlanta, Ga: American Cancer Society, 2015.
5. von Mehren M, Randall RL, Benjamin RS, et al. Soft Tissue sarcoma, version 2.2014 Featured updates to the NCCN Guidelines. J Natl Compr Canc Netw. 2012;10:951-60.
6. Ebaugh JL, Yuan M. Intimal sarcoma of the superficial femoral artery with osteosarcomatous differentiation. J Vasc Surg. 2010;53:1394-7.
7. Burke AP, Virmani R. Sarcomas of the great vessels. A clinicopathologic study. Cancer. 1993;71:1761-73.
8. Pérez del Río MJ, Molina Suárez R, Fresno Forcelledo MF, et al. Pulmonary artery intimal sarcoma. Immunohistochemical findings. Rev Esp Cardiol. 1998;51:850-2
9. Wong HH, Gounaris L, McCormack A, et al. Presentation and management of pulmonary artery sarcoma. Clin Sarcoma Res. 2015;5:3.
10. Yamamoto K, Nozue T, Tsuchida M, et al. Pulmonary embolism caused by intimal sarcoma of the pulmonay artery. Intern Med. 2012;51:3031-4.
11. National Cancer Institute: adult soft tissue sarcoma treatment (PDQ).
12. Sebenik M, Ricci A, DiPasquale B, Mody K. Undifferentiated intimal sarcoma of large systemic blood vessels. Report of 14 cases with immunohistochemical profile and review of the Literature. Am J Surg Pathol. 2005;29:1184-93.