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Treatment of intimal sarcoma of peripheral veins

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ABSTRACT

INTRODUCTION: Intimal sarcoma is an extremely rare group of undifferentiated pleomorphic sarcoma arising from the intimal layer of vessels accounting for only 1% of all sarcomas, intimal sarcoma of large veins are even less common.
CASES PRESENTATION: We present two cases of intima sarcoma, one originated form the basilar vein and the other from the cephalic vein, the first one was treated with surgery and postoperative chemotherapy followed by Radiotherapy (RT), the second case was treated with isolated limb perfusion followed by marginal resection and RT. Both patients progressed to the lungs in a short time, the first case was treated with metastasectomy of the lung and is without evidence of disease 7 months after surgery; the second case treated with isolated limb perfusion has stable disease.
DISCUSSION: Intimal sarcoma are very aggressive tumors, with a high metastatic potential, the two patients progressed to lung in a short time (2 months) after local treatment. Both cases exhibit good response to chemotherapy and metastasectomy with a disease – free period of 7 months.
CONCLUSION: We propose that given the aggressive behavior of these tumors, they should be treated with chemoradiotherapy postoperative, either by systemic chemotherapy or isolated limb perfusion for the limp sparing surgery in this histology.

1. Introduction

This work has been reported in line with the SCARE criteria [30]. Intimal sarcoma (IS) is defined as a malignant tumor arising in the tunica intima of large blood vessels. In the systemic circulation, most of IS derived from the aorta artery, accounting for only 1% of all sarcomas [1]. The primary neoplasm of the major blood vessels is divided into three categories based on their site of origin: a) From large veins (extremely rare group) b) Pulmonary artery and c) Aorta and its branches.

Large vessel intimal sarcomas tend to occur in the elderly with a history of peripheral vascular disease, and usually present with advanced disease with an aggressive course. The differential diagnosis of Intimal sarcomas should include benign lesions as well other soft tissue sarcoma (STS), diagnostic approach should have a high degree of suspicious, clinical manifestations are unspecific. Imaging should include an adequate and high-quality studies such as a Magnetic resonance imaging (MRI) preoperative to determine resectability as well as a CT scan of the chest, abdomen and pelvis to exclude metastatic disease [2–8].

Vascular neoplasms are classified as intimal sarcomas, angiosarcomas and leiomyiosarcomas; this last one represent most of the peripheral arterial tumors; To differentiate intimal sarcomas from the other two, histology and immunohistochemistry is needed. 26% of intimal sarcomas are well differentiated.

The cornerstone of treatment is extrapolated of STS, surgery with margins greater than 1 cm, limb-sparing surgery with RT are the standard of treatment [11,12]. For Stage II and III STS, preoperative chemoradiation, chemotherapy alone or with hyperthermia, postoperative chemotherapy are options but not the standard of treatment (as the literature shows not benefit). Preoperative RT increase wound complications and postoperative RT improves local control but not overall survival [16–23]. For unresectable disease the options of treatment include RT alone, chemoradiation or chemotherapy alone with the same considerations; tumors that become resectable can be treated with surgery follow by RT with or without chemotherapy [24]. Regional limb therapy has been evaluated with good results to preserve the limp, tumor necrosis factor – alpha and melphalan appear to have the best results [25–29].

There is very limited data available in the literature on surveillance strategies for STS, physical examination is the most important factor. No data exists in the follow up either with MRI, CT scans or

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Ultrasound. The median survival time is only a few months; aggressive tumors that can metastasize, the life expectancy is usually about 12–18 months [9].

2. Clinical case 1

A 61-year-old man presented with progressive increase in left forearm, physical examination reveals a multilobular, heterogenous mass of 5 × 6 cm, in the middle third, without impaired mobility, no distal neurovascular compromise, corresponding with the cephalic vein in the forearm.

The CT scan showed only the presence of a small rounded hypodense mass located in diaphragmatic dome, hepatic segment VIII measuring 12 mm compatible with simple cyst.

An incisional biopsy of the forearm was obtained and reported as pleomorphic and spindle cell sarcoma, intermediate grade at least. The tumor cells stained strongly positive for vimentin, which is found in both benign and malignant mesenchymal tumors, but were negative for other immunohistochemical stains such as Desmin (smooth muscle origin), cytokeratin and S100 protein (to ruled out metastatic carcinoma and melanoma respectively). The absence of CD34 staining argued against sarcoma origin of endothelial cells such angiosarcoma and malignant endothelioma.

In a multidisciplinary session, it was decided to take the patient to isolated limb perfusion with melphalan, in order to reduce its size followed by limb sparing surgery. The surgery was performed without any complications and type II Wieberdink toxicity was recorded. The after-treatment response was evaluated three months later as stable disease per RECIST 1.1 criteria (Figs. 1 and 2). Surgical procedure was done with a tumor marginal resection. The specimen consisted as an irregular vein, with overall dimensions of 20.1 × 5.9 × 4.2 cm. The final diagnosis was a high degree pleomorphic spindle cell sarcoma with cartographic necrosis that affects the whole course of the vessels compatible with intimal sarcoma. The immunohistochemistry was positive for vimentin and negative for S-100, CD56, MDM2 and CD34 (Figs. 3 and 4).

It was decided to continue with adjuvant radiotherapy completing a dose of 66 Gy in 33 fractions. 2 months after the end of adjuvant radiotherapy, the presence of multiple bilateral pulmonary nodules was documented, the largest with a diameter of

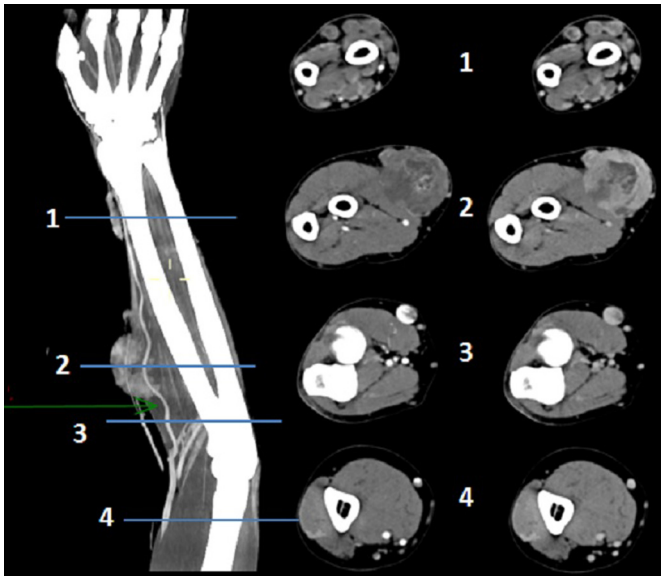

Fig. 1. CT scan, where different axial sections are shown, the presence of mass following superficial venous paths corresponding with the cephalic vein in the left forearm. The green arrow indicates the path of the radial artery.

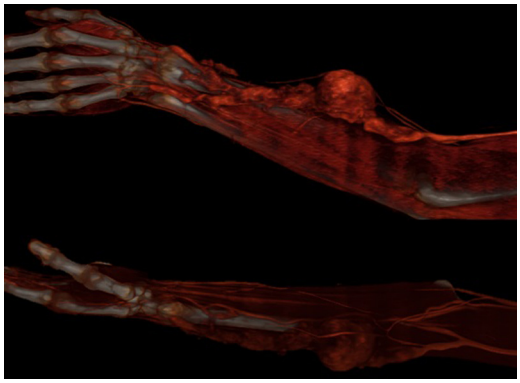

Fig. 2. 3D reconstruction where observe the presence of infiltration across the path of the cephalic vein.

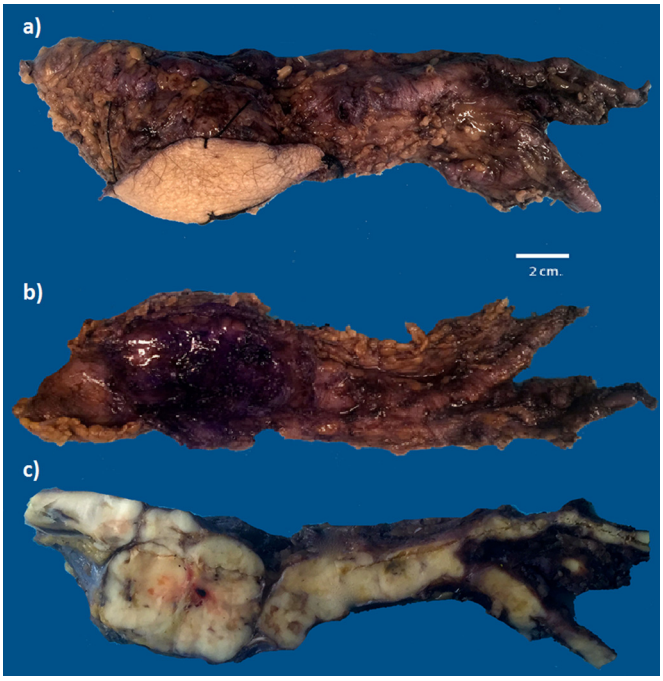

Fig. 3. Surgical specimen after marginal resección, the proximal edge is left and distal edge at the right. a) Anterior edge, b) surgical bed c) longitudinal section.

31 mm in the right parahiliar side. He received 6 cycles of doxorubicin as palliative chemotherapy. Currently the patient is without local recurrence, with stable lung metastatic disease and a 100% Karnofsky score, ECOG 0 and the limb with full functionality.

3. Clinical case 2

A 29 years old male diagnosed with Hodgkin lymphoma in 2004 treated with first- and second-line chemotherapy regimens, who achieved a complete response with no evidence of disease. In 2014 he was diagnosed with deep vein thrombosis secondary to a tumor in the left arm, Imaging studies reported a tumor in the medial compartment and middle third of the left arm apparently originated from the basilica vein, about 5 × 3 cm in its greater axes (Fig. 5). Core Needle biopsy reported a high-grade sarcoma with pleomorphic areas and production of osteoid material compatible with an intimal sarcoma.

Surgery was the first line of treatment, the findings were a left arm tumor, in the proximal third, 5 × 4 cm, subfascial, infiltrating the biceps muscle medially, with abundant newly formed vessels,