



Cooperation of Angiosurgeon and Orthopedist in a patient with intimal angiosarcoma in the left groin

Jana POBEHOVÁ, MD, Martina ZAVACKÁ, MD

Eastern Slovakia Institute for Cardiovascular diseases, P.J.Šafárik University, Medical Faculty, Košice, SLOVAKIA



INTRODUCTION

Intimal sarcoma (InS) is an extremely rare, mesenchymal neoplasm originating from large blood vessels and the heart, and it is one of the most common primary cardiac histologies /1,2/. Regarded as a high-grade tumor, it is marked by MDM2 nuclear overexpression and amplification of the 12q12-15 region (containing CDK4 and MDM2) /3/. These molecular features suggest that this pathway might play a relevant role in tumor pathogenesis. The outcome for InS patients is poor, with a reported median overall survival (mOS) in the range of 8 to 13 months /4,5/.

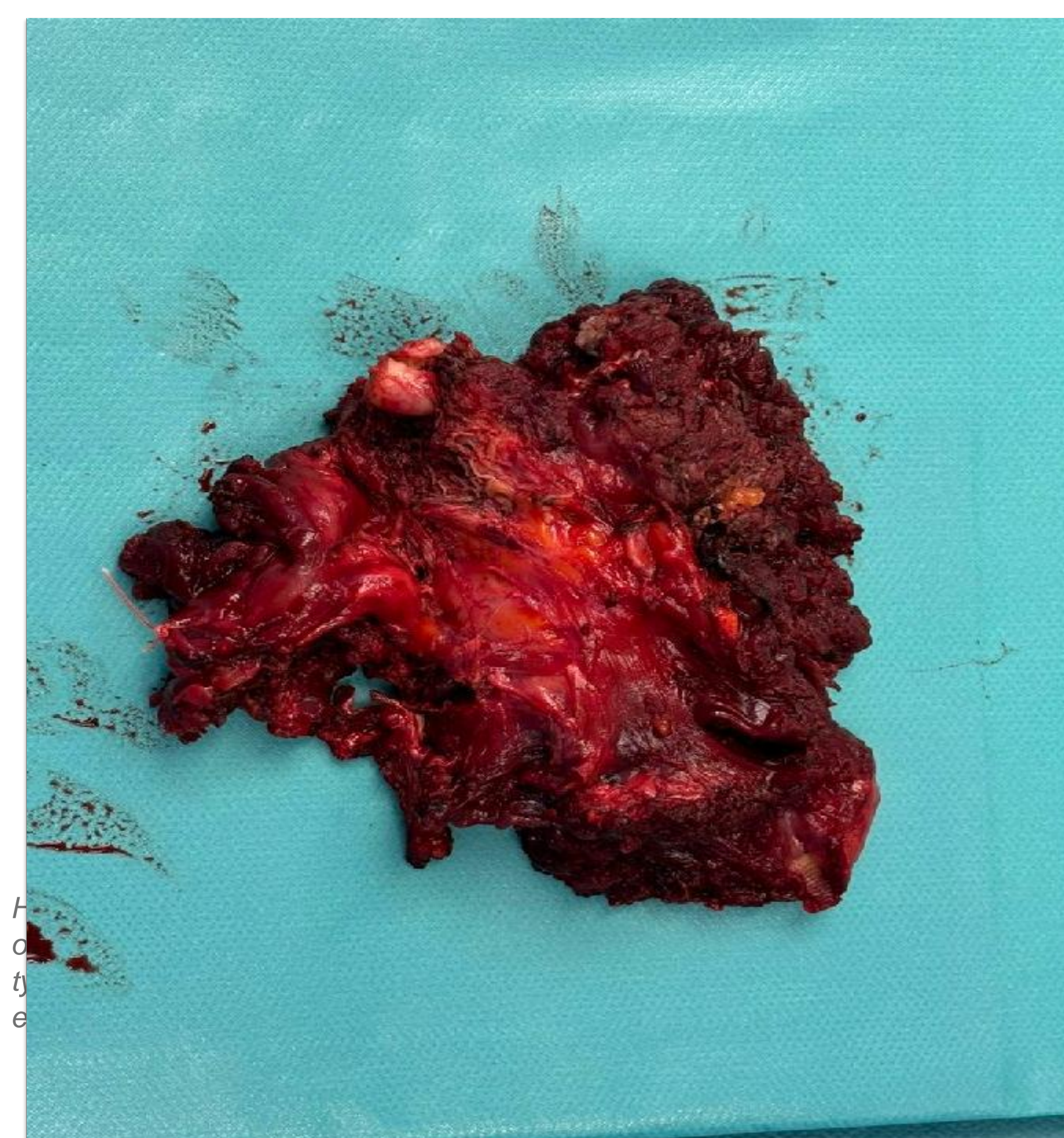
AIM

Intimal sarcoma is a rare neoplasm with an unfavorable prognosis. We present a case of a patient with a recurrence of intimate sarcoma in the area of the left groin, its recurrence and extensive reoperation, in which it was removed „in toto“.

Fig.1: surgical field after removal of a tumor in the groin



Fig.2: the first part of the excised tumor



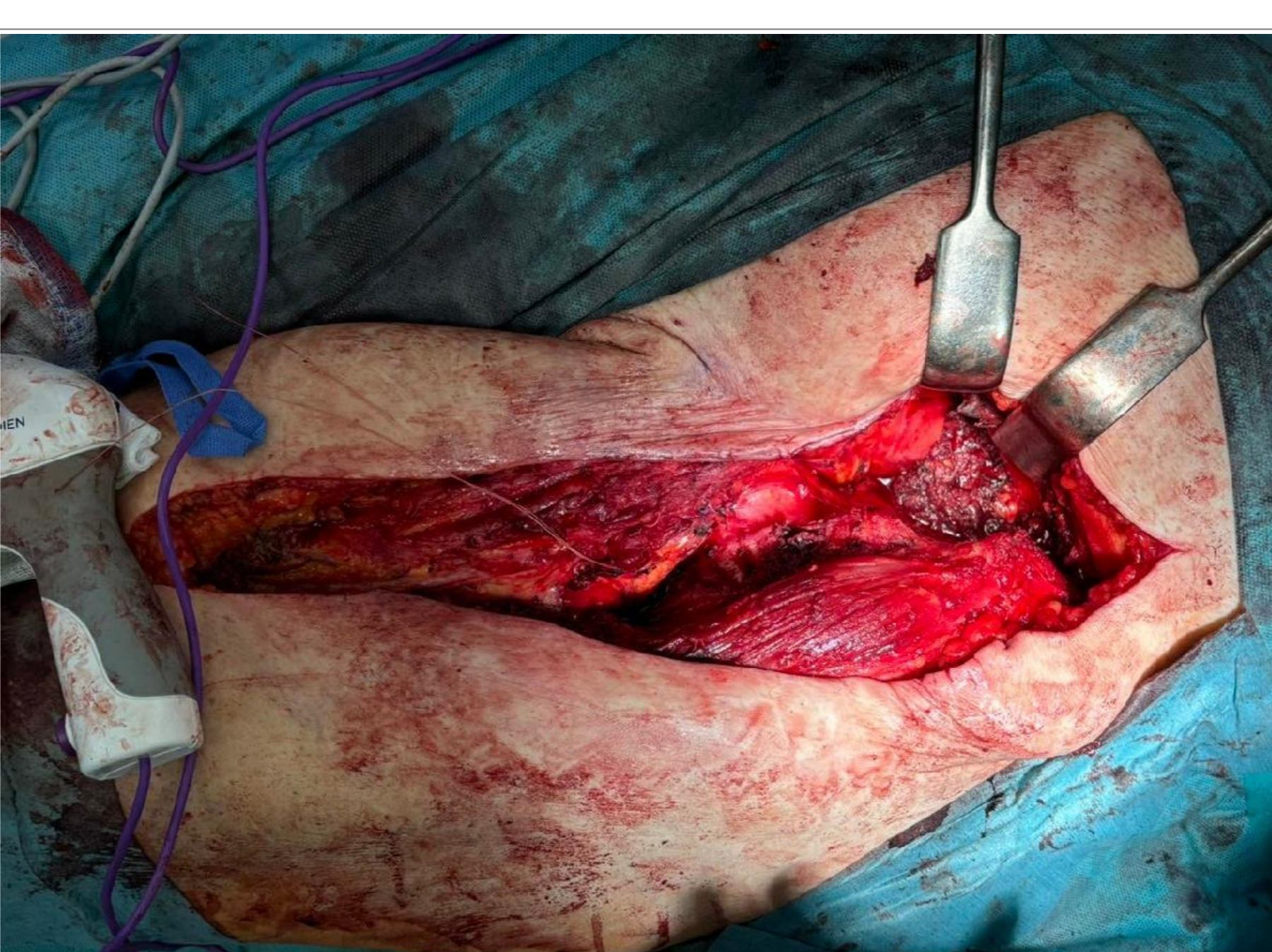
METHODS

We present a case of a 68-year-old patient with recurrence of angiosarcoma around the arteries of the left groin (AFP). He was primarily removed from his left groin in 2008 by liposarcoma, followed by radiotherapy. In 2023, there was a recurrence and overgrowth of the tumor in the area of the femoral arteries arteria profunda femoris /AFP/ on the left. We extirpated the tumor in its entirety, with a histological finding of high grade intimal angiosarcoma. Due to the exhaustion of radiotherapy options as the primary oncological treatment, a recurrence occurred within 1 year, this time affecting the anterior muscle group of the thigh/32x25x43 mm/. The patient began to have difficulty with the mobility of the limb. However, she was not at risk of ischemia, given that the recurrence did not affect the arteries of the thigh. After completing the CTAG and MRI examinations, the patient was operated on in cooperation with an orthopedist. The tumor was removed in its entirety, with negative edges. The wound gradually healed. The mobility of the left lower limb was preserved.

Fig.3: the second part of the excised tumor



Fig.4: condition after groin reconstruction



RESULTS

InS behave highly aggressive with a mean patients' survival ranging from 5 to 18 months /6/. Thus, patients are often diagnosed in an advanced disease stage. Furthermore, ISAs are often reported to be resistant to conventional chemotherapy /7/. Surgical treatment is a key part of these oncological diseases. Primary surgical treatment is the maximum possible tumor resection with a sufficient margin of safety tissues. However, the radicality of the performance must be taken into account possible mutilation of the patient (amputation of the tiny, pelvic exenteration). Radiotherapy, as well as surgical treatment represents an important treatment modalit as part of local treatment and disease control. The indication of radiotherapy has its limitations in particular in younger patients. When planning it, it is necessary to take into account possible late consequences such as are e.g. affecting growth, secondary malignancy in the irradiated field and others. Chemotherapy is a curative method. Low-risk patients can be treated less intensive treatment.

CONCLUSIONS

Management includes a multimodal approach /e.g. angiosurgeon, orthopedist, radiologist, oncologist, pathologist/ consisting of systemic chemotherapy, local surgery and radiotherapy.

Fig.5: the Angiosurgical team



BIBLIOGRAPHY

- 1 Fletcher CDM, Bridge JA, Hogendoorn PCW, Mertens F. WHO Classification of Tumours of Soft Tissue and Bone. Lyon, France: International Agency for Research on Cancer, 2013.
- 2 Neuville A, Collin F, Bruneval P, et al. Intimal sarcoma is the most frequent primary cardiac sarcoma: clinicopathologic and molecular retrospective analysis of 100 primary cardiac sarcomas. *Am J Surg Pathol.* 2014; 38: 461-469.
- 3 Bode-Lesniewska B, Zhao J, Speel EJ, et al. Gains of 12q13-14 and overexpression of mdm2 are frequent findings in intimal sarcomas of the pulmonary artery. *Virchows Arch.* 2001; 438: 57-65.
- 4 Van Dievel J, Sciort R, Delcroix M, et al. Single-center experience with intimal sarcoma, an ultra-orphan, commonly fatal mesenchymal malignancy. *Oncol Res Treat.* 2017; 40: 353-359.
- 5 Kato W, Usui A, Oshima H, Suzuki C, Kato K, Ueda Y. Primary aortic intimal sarcoma. *Gen Thorac Cardiovasc Surg.* 2008; 56: 236-238.
- 6 WHO Classification of Tumours of Soft Tissue and Bone, 5th edn, Vol. 3 (2020).
- 7 Frezza AM, Assi T, Lo Vullo S, Ben-Ami E, Dufresne A, Yonemori K, et al. Systemic treatments in MDM2 positive intimal sarcoma: a multicentre experience with anthracycline, gemcitabine, and pazopanib within the World Sarcoma Network. *Cancer.* 2020;126:98-104.
- 8 Staats P, Tavora F, Burke AP. Intimal sarcomas of the aorta and iliofemoral arteries: a clinicopathological study of 26 cases. *Pathology.* 2014;46:596-603.
- 9 Burke AP, Virmani R. Sarcomas of the great vessels. A clinicopathologic study. *Cancer.* 1993;71:1761-73.
- 10 Tavora F, Miettinen M, Fanburg-Smith J, Franks TJ, Burke A. Pulmonary artery sarcoma: a histologic and follow-up study with emphasis on a subset of low-grade myofibroblastic sarcomas with a good long-term follow-up. *Am J Surg Pathol.* 2008;32:1751-61.
- 11 Mandelstamm M. Über primäre Neubildungen des Herzens. *Virchows Arch.* 1923;43-54.
- 12 Hottenrott G, Mentzel T, Peters A, Schroder A, Katenkamp D. Intravascular ("intimal") epithelioid angiosarcoma: clinicopathological and immunohistochemical analysis of three cases. *Virchows Arch.* 1999;435:473-478.
- 13 Staats P, Tavora F, Burke AP. Intimal sarcomas of the aorta and iliofemoral arteries: a clinicopathological study of 26 cases. *Pathology.* 2014;46:596-603.