



Long-term survival after resection of a primary leiomyosarcoma of the innominate vein

Report of a case

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Leiomyosarcoma of the innominate vein is a rare but usually lethal disease. We report the case of a 50-year-old woman, undergoing a curative resection of the tumor. She is alive and free of disease at 88-month follow-up. Surgical excision remains the current optimal treatment able to provide a chance of cure.

KEY WORDS: Late survival, Venous leiomyosarcoma

Introduction

Primary leiomyosarcoma of the innominate vein is a rare disease whose prognosis is usually dismal. Nonetheless, two isolated cases of long-term survival after excision of the neoplasm have been reported so far ^{1,2}. We now report a third case of late survival after curative resection of this infrequent condition.

Case Report

A 50-year old woman was referred for a soft, left latero-cervical, 2 cm diameter mass which resulted to be undifferentiated carcinoma at excisional biopsy. A CT-

scan showed a mass extending from the left internal jugular vein to the innominate vein (Fig. 1, which was confirmed as an intravascular tumor at Positron Emission Tomography). On June 2005 the tumor was resected "en bloc" with the internal jugular and innominate vein. A radical neck and supra-clavicular lymphadenectomy was also performed. Histology was consistent with an intravascular epithelioid leiomyosarcoma with micrometastases in two mediastinal lymph nodes. The clinical and surgical details of the case have been previously reported ³. The patients made an uneventful recovery. Radiation and chemotherapy were not performed. One year later the patient presented a lymphatic edema of the left upper limb, probably caused by lymphadenectomy, which could be managed with physical therapy. She undergoes regular controls on a yearly basis at the Department of Oncology and remains alive and free from disease at 88-month follow-up.

Discussion

Venous leiomyosarcomas are slowly growing tumors affecting most commonly the Inferior Vena Cava ⁴⁻⁶. In other primary localizations they are extremely rare ⁷. Due to their slow growing pattern they usually allow a suf-

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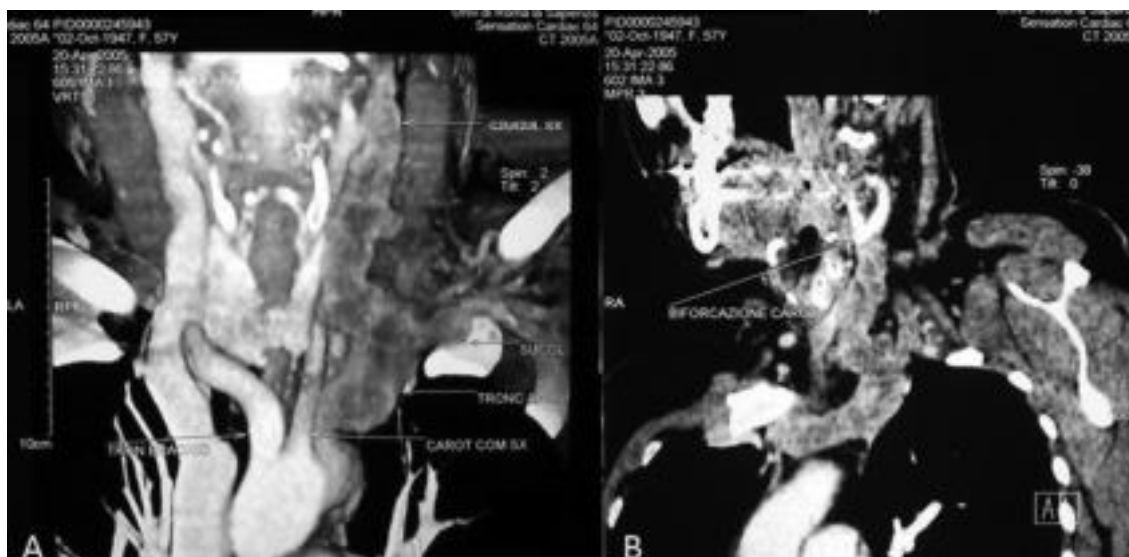


Fig. 1: Helical CT scan of the neck and upper chest. Extensive intravascular neoplastic growth in the internal jugular – innominate vein (A) with extravascular extension toward the carotid bifurcation (B).

ficient collateral flow to develop, therefore they rarely cause symptoms related to venous thrombosis, and metastatic spread, usually to the lungs, is usually evident within two years from the diagnosis of the primary tumor. Late results of surgical excision and late survival are usually very disappointing, as these tumors are often fairly resistant to radiation and chemotherapy. Nonetheless, sporadic cases of long-term survival after excision of venous leiomyosarcomas have been reported. In particular, concerning leiomyosarcomas of the internal jugular – innominate vein, to our knowledge, only two previous cases of survival beyond 5 years, after surgical excision have been reported so far ^{1,2}. In both cases long-term survival was obtained with a radical surgical excision of the primary tumor. Our case supports these earlier findings and underscores the importance of radical surgical excision and lymphadenectomy as the only actual mean to obtain a potential long-term survival. In the reported patient the extended lymphadenectomy had a sequela consisting of edema of the left upper limb, which caused mid discomfort, but can be controlled with periodical physical therapy. Nonetheless, an extended lymphadenectomy was mandatory for an effective protection from potential local and distal recurrences. Finally, the obtention of this satisfactory result without adding any adjuvant therapy underscores the limited value and efficacy of radiation and chemotherapy in this setting, due to the resistance of sarcomas in general to adjuvant treatments. In conclusion, the reported case supports the assumption that, although unfrequent, long-term survival after surgery for rare and potentially lethal conditions, as leiomyosarcomas of the innominate vein is possible. Aggressive surgical treatment should always be attempted whenever possible, in order to offer patients the only chance of cure.

Riassunto

Il leiomiomasarcoma della vena anonima è una condizione rara a prognosi quasi sempre infausta. Si descrive il caso di una donna di 50 anni operata di resezione curativa per questa patologia. La paziente è in buona salute e libera a recidive a 88 mesi dall'intervento. Il trattamento chirurgico rimane il trattamento di scelta per una possibile risoluzione duratura di questa neoplasia.

References

1. Miller MM, Munnel ER, Poston A, Harkey MR, Gratham RN: *Primary angiosarcoma of the innominate vein: a case report with resection and long-term survival*. J Thorac Cardiovasc Surg, 1985; 90:148-50.
2. Rueckert RI, Kroencke TJ, Buerger K: *Long-term survival after radical resection of a primary angiosarcoma of the innominate vein*. Ann Thorac Surg, 2000; 70:1713-714.
3. Illuminati G, Miraldi F, Mazzesi G, D'urso A, Ceccanei G, Bezzi M: *Primary leiomyosarcoma of the innominate vein*. Ann Vasc Surg, 2007; 21(1):75-78.
4. Illuminati G, Calio' FG, D'Urso A, Giacobbi D, Papaspyropoulos V, Ceccanei G: *Prosthetic replacement of the infra-hepatic inferior vena cava for leiomyosarcoma*. Arch Surg, 2006; 141(9):919-24; discussion 924.
5. Hines OJ, Nelson S, Quinones-Baldrich WJ, Eilber FR: *Leiomyosarcoma of the inferior vena cava: Prognosis and comparison with leiomyosarcoma of other anatomic sites*. Cancer, 1999; 85(5):1077-83.
6. Kieffer E, Berrod JL, Chomette G: *Primary tumors of the inferior vena cava*. In: Bergan JJ, Yao JST, editors. *Surgery of the Veins*. Orlando, FL: Grune & Stratton, 1985; 423-43.
7. Dzsinić C, Gloviczki P, van Heerden JA, Nagorney DM, Pairolero PC, Johnson CM, Hallett JW Jr, Bower TC, Cherry KJ Jr: *Primary venous leiomyosarcoma: A rare but lethal disease*. J Vasc Surg, 1992; 15(4):595-603.