An unusual tumor in a rare localization: intravascular leiomyosarcoma of the cephalic vein.

Case report

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Vascular leiomyosarcoma is a very rare soft tissue neoplasma, with a poor prognosis. We report a vascular leiomyosarcoma arising from an unusual site: the media-vascular wall of the cephalic vein. A 71-year-old man with a subcutaneous node on the volar side of right forearm, came to our attention with signs of vascular obstruction and venous stases, without functional or sensory loss of the right upper extremity. We successfully performed a surgical excision of the cancer with a wide margin resection. During surgery no axillary lymphnodes metastes were found. 4 months after surgery the wound had fully healed, and he suffered no effects by surgery, but he became lost to long-term follow-up and refused adjuvant radiotherapy or chemotherapy. This case report shows that, in presence of venous stases or thrombosis signs of the upper extremity, leiomyosarcoma of the cephalic vein must be considered in the differential diagnosis among lung cancer, lymphoma or mesenchymal tumours.

KEY WORDS: Chepalic vein, Leiomyosarcoma

Introduction

Vascular leiomyosarcoma is a rare neoplasma, it can be divided into three types according to their site of origin: 1) Leiomyosarcoma of soft tissues; 2) cutaneous leiomyosarcoma; 3) vascular leiomyosarcoma. On vascular leiomyosarcomas there are few publications about clinical presentation and management. They are 5 times more frequent in veins than in arteries and arise directly from the smooth muscular cells. In the literature are reported many cases in big vessels (caval vein, iliac vein and others)\(^1\), but it is uncommon to find such neoplasma in smaller vessels as we reported \(^2\)-\(^3\). The prognosis is uncertain \(^{1,4,5}\) and currently the majority of cases are treated like a sarcoma.

Case report

A 71-year-old man presented in our clinic showing a painful, swelling node on volar side of the right forearm and shaped in the last year increased in size. On clinical examination we reported a solid swelling of about 2 cm with signs of vascular obstruction and venous stases, without functional or sensory loss of the right upper extremity. The lesion was mobile respect to the skin and fixed to deeper tissue. The anamnesis was negative, and physical examination did not show any other disorders for limphonode masses in the axilla.
An intralesional biopsy was performed. The relative pattern was spindle cells tumor with eosinophilic cytoplasm and cigar shaped nuclei and anisocariosis with a suspected diagnosis of a leiomuscolar neoplasia, therefore a complete excisional biopsy was planned preserving the soft tissue around.

During the surgery (Fig. 3) was observed that the leiomyosarcoma was responsible of subocclusion of the cephalic vein. T umor exposure was obtained via a linear incision.

The histological diagnosis confirmed the previous biopsy: the neoplasm was vascular leiomyosarcoma with poly-ploid endovasal growth with cytologic atypias and high mitotic rate (9-10 x 10 HPF). It originated from the media of the cephalic vein, growing and infiltrating extra-vascular surrounding soft tissues. Immunohistochemically the majority of tumor cells were positive for desmin, smooth muscle actin. There was a negativity reaction for S.100 and a CD 31 positivity only for endothelial cells.

Final diagnosis of the excised tumour was a leiomyosarcoma grade II with a diameter of 2.5 cm radically excised.

After the surgery the patient refused any adjuvant radiotherapy or chemiotherapy.

Discussion and conclusion

Leiomyosarcoma only account for 5-10% of malignant soft tissue tumours in adults 6. Four main locations for leiomyosarcoma origin can be distinguished: 1)intrabdominal/retroperitoneal; 2)cutaneous; 3)subcutaneous; 4)vascular, the most rare. Vascular leiomyosarcoma is a rare neoplasma that more frequently occurs in the retroperitoneal space, affecting in 75% the inferior cava vein7.

Ultrasounds images revealed a lobulated hypoechogenic mass. An additional contrast enhanced MRI of the right forearm (Fig. 1) confirmed the presence of a nodular solid neoplasma 2.5 cm in diameter, localized in proximity of brachioradialis muscle and Carpal radial flexor muscle.

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Few cases involving venous branches of the lower extremity are reported in literature. Whereas only single-case-reports show upper extremity tumour manifestation, furthermore head and neck region and azygos vein were found. The majority of venous leiomyosarcomas involve in large-to-medium sized vessels (41%), 38% in the inferior cava vein, 12% in the pulmonary artery, 9% in the systemic arteries. Leiomyosarcoma deriving from smaller vessels are an exception which may lead to nervous or arterial compression and often protrude through small lumina of adjacent venous branches. Moreover leiomyosarcoma of thin veins, extended from the tunica media, early infiltrates into surrounding soft tissue. Biopsy diagnosis of soft tissue leiomyosarcoma is always necessary to consider the rare possibility of a primary intravascular tumour growth. Histomorphology, immunohistochemical staining for smooth muscle markers facilitates the correct diagnosis. Leiomyosarcomas of a vascular origin appear to be associated with a more aggressive tumour growth and with a poorer prognosis if compared to respective cancers of the soft tissue and requires adjuvant radiation therapy. In a large number of cases vascular-leiomyosarcoma is diagnosed after the cancer has already progressed to distant areas. Microthrombembolism and pulmonary metastases may complicate the course of the disease. In this report, we have presented a rare case of intravascular leiomyosarcoma in the uncommon anatomical site of the cephalic vein. Actually prognosis in this type of tumor is poor, with frequent pulmonary metastasis and adjuvant chemio or radio-therapy do not significantly increase survival rate.

Riassunto

Un tumore inusuale, in una localizzazione rara: leiomiosarcoma intravascolare della vena cefalica, caso clinico.

I leiomiosarcomi con origine da vasi di piccolo calibro sono molto rari e poco documentati in letteratura, in modo particolare è rara la localizzazione a livello dell’arto superiore. In questo caso clinico viene presentato un leiomiosarcoma intravascolare della vena cefalica, senza localizzazione metastatica linfonodale, clinicamente evidente come nodulo dolente sottocutaneo e con segni di ostruzione vascolare e stasi venosa a carico dell’arto superiore interessato.

References
