Case report

Myxoid liposarcoma of the hand – A rare entity

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ABSTRACT

Hand liposarcomas are not well recognized in common practice, which leads to erroneous diagnostics and delayed proper treatment.

We reported a case of a myxoid liposarcoma classified as stage IIb, and developed in the palmar side of the left hand in a 38-year-old man. There was an initial tumor reduction by a sclerosing agent injection that proved to be a failure. An open excisional biopsy confirmed the diagnostic after pathology examination. Recurrence occurred after 2 years, followed by a second resection and an adjunctive radiotherapy. At 5 years follow-up, the patient was recurrence free.

Although myxoid liposarcoma is rare in the hand, it should be considered in the differential diagnosis of a painless soft tissue mass in this region.

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1. Introduction

Hand liposarcomas (LS) are not well recognized in common practice, which leads to erroneous diagnostics and delayed proper treatment. Myxoid liposarcoma (MLS) is the most common type of LS and is known to have a greater affinity to the lower limb.¹ Its occurrence in the upper limb, and especially in the hand, seems to represent a rare entity.² As shown in our singular observation, neoplastic etiology must be systematically ruled out in case of any subcutaneous lesion.

2. Case presentation

We reported the case of a 38-year-old, left hand dominant man, with history of a workplace accident, 18 years ago, causing multiple deep wounds of the dorsum of the left hand, with no sensory or motor deficit. The patient has a 1-year history of a painless, slow-growing mass in the palm of the left hand with occasional hot flushing of the thumb. Physical examination revealed a palmar tender painless immobile mass in the thenar eminence, measuring approximately 1.5 cm × 1 cm (Fig. 1).

A plain radiograph of the left hand showed a soft tissue mass in the form of increased soft tissue density (Fig. 2). A soft tissues ultrasound was performed, showing a vascular echogenic tissular mass of 1.9 cm of the major axis in contact with the ulnar thumb pedicle.

Open biopsy was indicated. Taking into consideration the vascular relations of the tumor, the procedure was preceded by an attempt of tumor reduction by embolization during angiography (Fig. 3). After a short follow-up period of 6 weeks, the swelling increased rapidly reaching 5 cm of the long axis with a slight inflammatory reaction.

The magnetic resonance imaging (MRI) was not available during that time, with a risk of delaying tumor management. We performed a computed tomography (CT) scan that revealed a low-density, heterogeneous, poorly delimited lesion developed in the palm of the left hand, measuring 7 cm × 3 cm, with contrast enhancement after intravenous injection and no sign of bone involvement or calcifications (Fig. 4). We completed the investigation by a thoraco-abdomino-pelvic CT scan with no signs of secondary localization of the tumor.

We performed a surgical excisional biopsy after a 2 months' delay from embolization. Large tumoral excision was done sacrificing the ulnar thumb pedicle. Gross examination found a poly-lobed, greyish and friable tumor. Histology confirmed the diagnosis of a MLS. No round cell component was seen. The margins of excision were wide added to a tumor-free on the macroscopic and microscopic examination. No adjuvant treatment was done.

The tumor was classified as stage IIb, and based on The American Joint Committee on Cancer (AJCC) staging system of STS.³

Two years after surgery, the evolution was characterized by the recurrence of the left hand mass, measuring 5 cm × 3 cm, with no general extension. The patient underwent a second surgical excisional biopsy taking away the ancient scar and the tumoral mass; index flexors and radial pedicle were not affected. Excision

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Fig. 1. Left hand photo showing a thenar's lodge mass at first examination.

Fig. 2. Preoperative AP view left hand X-ray, showing an increased soft tissue density in the thenar's lodge.

Fig. 3. Left hand angiography showing a vascularized mass repulsing surrounding blood vessels.

Fig. 4. A preoperative CT scan showing a suspicious vascularized mass of the palm of the hand.

Fig. 5. Histologic boards concluding to a myxoid liposarcoma.

margins were tumor-free but marginal. Surgery was followed by post-operative radiotherapy with a total dose of 54 Gy, spread over 5 weeks. The second histologic examination reaffirmed the recurrence of the MLS (Fig. 5).

At a five years follow-up, no local tumoral recurrence and no signs of systemic invasion were found. The overall functional rating was 83% according to the International Society of Limb Salvage rating system.

3. Discussion

Sarcomas originate from primitive mesenchymal cells. They account for 0.1–0.2% of all cases of adult's neoplasms. Approximately 20% of malignant STS are represented by LS.⁴ Numerous reports of LS have been documented in the literature with an estimated incidence of 2.5 case/million/year.²

Malignant tumors of the hand are uncommon. They represent 2–5% of all hand tumors.¹ The MLS is an even more rare entity in the hand. To the best of our knowledge, only one case has been reported in the literature.⁵ The etiology of most tumors of the hand is unclear. Heredity, metabolic disturbances, and trauma have been cited as significant factors. In the reported case, ancient hand trauma may represent the inducing factor for the appearance of MLS.

Extremity lesions are generally sampled by multiple core needle biopsies to identify the histological subtype and to stage the disease. Open biopsy is still indicated but with a higher morbidity. In the reported case, an attempt of tumor reduction by embolization was prior to biopsy; this was based on the vascular nature of the mass on ultrasounds. Given the failure of this attempt, fine needle biopsy should have been done in the first place.

Nowadays huge progresses have been achieved in limb salvage surgery techniques and adjuvant therapy. Limb-sparing surgery

with or without adjuvant RT is an effective treatment option for extremity STS.⁶ Different studies have suggested that MLS tends to be relatively more radiosensitive than other types of STS, with high rates of regression and even complete clinical response.⁷

The National Comprehensive Cancer Network (NCCN) panel guidelines for stage II–III MLS are: For resectable tumors, surgery followed by radiotherapy is the primary treatment. If the tumor is unresectable, neoadjuvant therapies (radiotherapy, chemoradiation, chemotherapy) are recommended primarily, even if some authors have not found any improvement in overall survival.⁸ Tumors that become resectable after preoperative treatment can be treated with surgery. Otherwise, definitive radiotherapy is recommended.⁹ In the reported case, excisional biopsy without adjuvant treatment was done on first surgery. After tumoral recurrence, an adjuvant radiotherapy followed excisional surgery with a tumor-free follow-up period of 5 years. This led us to insist on the importance of adjuvant treatment in the management of such tumors.

4. Conclusions

MLS is a rather rare disease, especially when it involves the hand. Extra-pulmonary recurrence is relatively frequent. Correct preoperative diagnosis and subsequent appropriate treatment can achieve good oncologic and functional outcomes for patients with MLS of the hand. Adequate adjunctive therapies may help achieve low recurrence rates.

Competing interests

A written informed patient consent was obtained for the publication of this article.

Conflicts of interest

The authors have none to declare.

References

1. Thomas BP, Sasi K, Pallapati SCR, Mathew A, Sreekanth R, Thomas M. Malignant tumours of the hand and wrist. *Indian J Plast Surg.* 2011;44(2):337–347.
2. Henze J, Bauer S. Liposarcomas. *Hematol Oncol Clin North Am.* 2013;27(5):939–955.
3. Edge SB, Compton CC. The American Joint Committee on Cancer: the 7th edition of the AJCC cancer staging manual and the future of TNM. *Ann Surg Oncol.* 2010;17(June (6)):1471–1474.
4. Enzinger FM, Winslow DJ. Liposarcoma. A study of 103 cases. *Virchows Arch Pathol Anat Physiol Klin Med.* 1962;335:367–388.
5. Buehler D, Marburger TB, Billings SD. Primary subcutaneous myxoid liposarcoma: a clinicopathologic review of three cases with molecular confirmation and discussion of the differential diagnosis. *J Cutan Pathol.* 2014;41(December (12)):907–915.
6. Lin PP, Guzel VB, Pisters PW, et al. Surgical management of soft tissue sarcomas of the hand and foot. *Cancer.* 2002;95:852–861.
7. Chung PW, Dehesi BM, Ferguson PC, et al. Radiosensitivity translates into excellent local control in extremity myxoid liposarcoma: a comparison with other soft tissue sarcomas. *Cancer.* 2009;115(14):3254–3261.
8. Bramwell V, Rouesse J, Steward W, et al. Adjuvant CYVADIC chemotherapy for adult soft tissue sarcoma—reduced local recurrence but no improvement in survival: a study of the European Organization for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group. *J Clin Oncol.* 1994;12:1137–1149.
9. Demetri GD, Antonia S, Benjamin RS, Bui MM, Casper ES, Conrad III EU. Soft tissue sarcoma. *J Natl Compr Cancer Netw.* 2010;8:630–674.