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Olgu Sunumu / Case Report

Subcutaneous Reccurent Myxoid Liposarcoma

Subkutan Nükseden Miksoid Liposarkom

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Abstract

Liposarcoma, one of the most common soft-tissue sarcomas, originates from primitive mesenchymal cells, and its diagnostic have been well established. Myxoid liposarcoma is the second most common histological subtype, occurring more frequently during the fourth and fifth decades of life. We describe a case of recurrent myxoid liposarcoma of the right flank in 50 years old man. Treatment of the recurrence involved chemotherapy neo-adjuvant (3 courses), wide surgical resection (resection R1) followed by locoregional radiation therapy. Patient's evolution was marked by complete remission maintained after 12 months follow-up. The myxoid liposarcoma has a low rate of local failure with trimodality therapy combined chemotherapy, radiotherapy and surgery.

Keywords: Myxoid liposarcoma (MLS), recurrence, chemotherapy, radiotherapy, surgery

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En yaygın yumuşak doku sarkomlarından biri olan liposarkom, ilkel mezenkimal hücrelerden kaynaklanır ve tanısı iyi bilinmektedir. Miksoid liposarkom, yaşamın dördüncü ve beşinci dekatlarında daha sık görülen ikinci en yaygın histolojik alt tiptir. Biz 50 yaşında erkek hastada tekrarlayan miksoid liposarkom olgusunu tanımladık. Nüksün tedavisi neoadjuvan kemoterapi (3 kür), geniş cerrahi rezeksiyonu (rezeksiyon R1) ve lokalrejyonel radyasyon tedavisini takip etti. Hastanın gelişimi 12 aylık takipten sonra devam eden tam remisyon ile işaretlendi. Miksoid liposarkom, trimodalite tedavisi kombine kemoterapi, radyoterapi ve cerrahi ile düşük lokal yetmezliğe sahiptir.

Anahtar Kelimeler: Miksoid liposarkom (MLS), nüks, kemoterapi, radyoterapi, cerrahi

INTRODUCTION

Liposarcomas are soft tissue sarcomas arising from adipose tissue. In 2013, the World Health Organization updated their classifications for bone and soft tissue sarcomas to include three different subtypes of liposarcoma: well-differentiated or de-differentiated liposarcoma, pleomorphic liposarcoma, and myxoid/round-cell liposarcoma (1). These subtypes are based upon both clinicopathologic and molecular differences. Myxoid liposarcoma is the second most common subtype of liposarcoma.There is evidence to suggest that MLS is both radioresponsive and radiosensitive (2). Comparative analysis of MLS versus other sarcoma subtypes suggests greater response rates in MLS with addition of anthracyclines-based chemotherapy (3). However, there is relatively limited information on factors that predict for overall outcome in MLS, particularly with regard to the benefits of combined trimodality therapy.

CASE REPORT

A man 50 years old, with no pathological antecedents, who presented since 2011 a slowly growing mass of the right flank. Magnetic Resonance Imaging (MRI) showed an Inhomogeneous tumor of 6*8*9 cm3 in size suspicious for sarcoma. Surgery was recommended and the tumor was resected circumferentially. The anatomo-pathological study was in favor of grade I myxoid liposarcoma without round cell component (figure 1) .

The patient was lost for follow-up since 2012. three years later, the patient was referrerd to manage a giant tumefaction at the same initial site. The mass made more than 20cm at the clinical examination with inflammatory

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signs, fixed in relation to both planes and of firm consistency.

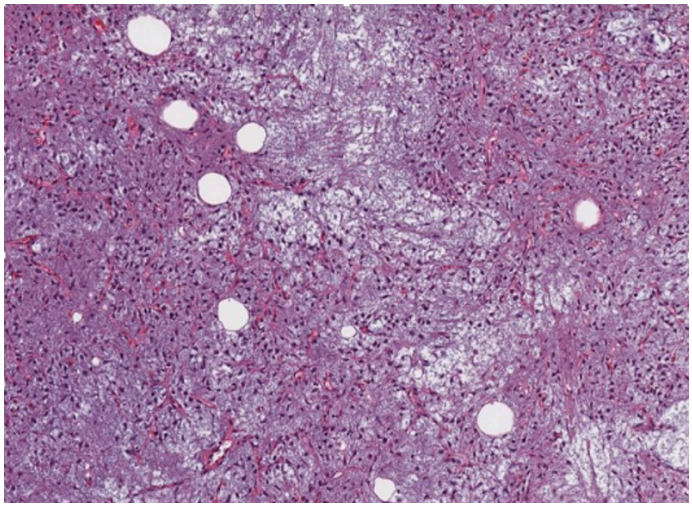

Figure 1. Microscopic pathology image showing Myxoid Liposarcoma

A thoracic and abdomino pelvic Computer Tomograph (CT) scan was realized, it shows a heterogeneous bilobular tissue mass of the superficial soft tissues next to the right iliac crest measuring 10*12 cm and extended to a height of 18.4 cm; there was no metastasis detected.

Due to bad prognosis criteria which are: Recurrence, rapid increase in the volume of the mass, the presence of inflammatory signs and the histological type (chemosensitive); the patient has received 3 courses of neo-adjuvant chemotherapy type MAI (Epirubicine, ifosfamide, G-CSF) with a good reponse (more than 50% clinical and radiological response). The patient benefited from a surgical exeresis; the histological aspect was in favor of a myxoid grade I liposarcoma according to French Federation of Cancer Centers Sarcoma Group (FNCLCC), presence of a round cell component estimated at 2%; the tumor was located 3 mm from the upper and lower limit, 8 mm from the internal limit, 15 and 10 mm from the lower and the outer limit respectively (resection R1). The surgical revision was impossible, and the decision was to complete with adjuvant radiotherapy after complete healing. The patient received loco-regional radiotherapy at the total dose of 66 Gy in 2 series: a dose of 50 Gy in 25 fractions of 2 Gy/Fr + a complement of 16 Gy in 8 fractions of 2 Gy/Fr (figure 2).

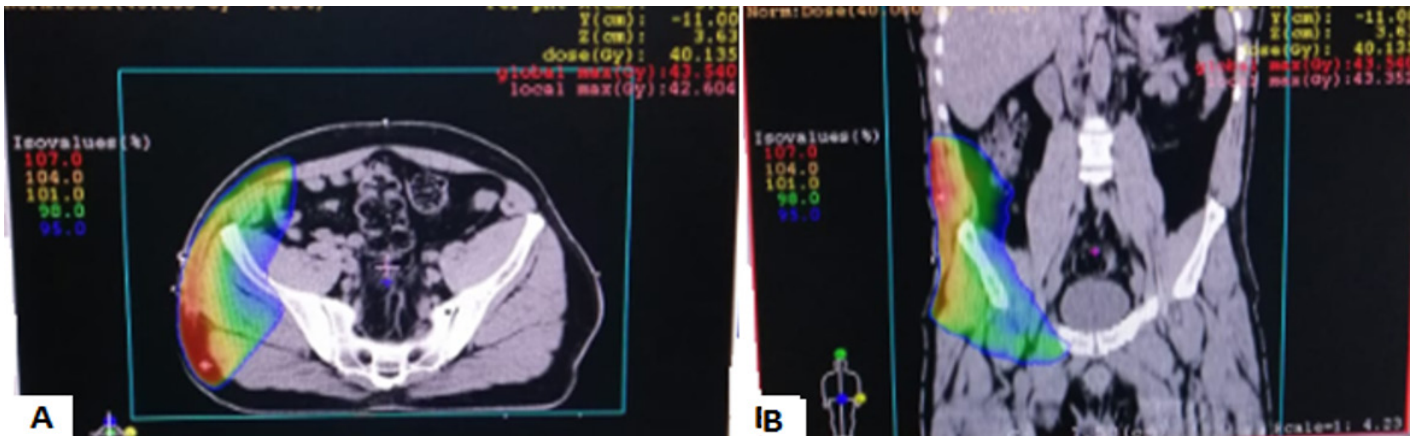

Figure 2. A= Axial tomodensitometric sections of 3 cm thick: Dose distribution of the patient's 3D radiation
B=Frontal tomodensitometric section : dose distribution of the patient's 3D radiation

The evolution was marked by a clinical and radiological remission complete maintained after a decline of 12 months.

DISCUSSION

Surgery remains the main treatment of MLS. However recent studies have shown that MLS tumors are sensitive to radiation therapy. Surgery, is therefore, often combined with neoadjuvant or adjuvant radiation and/or chemotherapy (4). The optimal goal of surgery is to successfully remove the tumor with a function sparing procedure maintains a wide excision with negative margins. The ability to perform a limb-sparing surgery is based on the extent to which important anatomical structures including fascial layers, blood vessels, nerve sheays, and muscle fibres are abutting or invaded by the tumor (5). However, while positive surgical margins lead to a higher risk for local recurrence, this has not been shown to correlate with an increased risk for metastatic disease or wore overall survival (6-7). In case where surgery it out of option, radiation and chemotherapy is employed.

MLPSs have been found to be much more radiosensitive than many other types of soft tissue tumors (8). A Retrospective review at M.D Anderson evaluated 112 patients with liposarcoma (71 of these patients had myxoid round cell) that were treated with conservative surgery and radiation with the goal to evaluate local control, metastatic relapse and overall survival. These patients had an excellent local control of over 90% at 10 years and the metastatic controversy as

to whether noadjuvant versus adjuvant radiation is more advantageous with regard to clinical outcomes. Therefore, given the lack of any proven difference in rates of local controle from pre-operative radiation, the standard of care has been to utilize adjuvant radiation unless it was determined that a limb-salvage surgery could not be completed due to the size of the tumor, and that tumor shrinkage could allow for resection without amputation (9).

The decision to give adjuvant chemotherapy is based on the risk of recurrence with metastatic disease. High-risk soft tissue sarcomas warrant consideration of adjuvant therapy. The benefit of adjuvant chemotherapy was demonstrated in the Italian sarcoma study Groups randomized comperative trials. This study demonstrated an increase in disease-free survival as well as median overall survival (10). More recently, an international multi-center randomized phase III trial was performed to determine the benefit of a histology-tailored neoadjuvant chemotherapy regimen in high grade soft tissue sarcomas. In this study, the patients enrolled with myxoid liposarcoma were randomized either to trabectedin or the combination of epirubicin and ifosfamide. No advantage to histology-tailored chemotherapy was observed in this study. In fact, there was a 20% difference in both 3 year disease-free and overall survival in favor of the standard chemotherapy group, offering further evidence for the utility of this regimen in the curative setting for all high-grade soft tissue sarcomas (11). In most instances, adjuvant chemotherapy is chosen over neo-adjuvant therapy given there is no evidence that one is superior in terms of overall survival.

CONCLUSION

Given the rarity of these tumors, myxoid liposarcomas similar to all sarcomas, are best evaluated by a multidisciplinary team, which includes recommendations from a radiologist, pathologist, surgeon, medical oncologist, and radiation oncologist with expertise in soft tissue sarcoma. Management of these tumors once appropriately identified and histologically confirmed, typically consists of multimodality treatment with surgery and radiation with or without chemotherapy.

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