REPORTS OF PRACTICAL ONCOLOGY AND RADIOTHERAPY 15 (2010) 107–109

available at www.sciencedirect.com

ScienceDirect

journal homepage: http://www.rpor.eu/

Case report

# Langerhans cell sarcoma: Response to radiotherapy

*Anna Lucas*[a,*], *Eva González Barca*[b], *Octavio Servitje*[c], *Lina Abenoza*[b], *María Noel González*[a], *Miquel Macià*[a], *Alicia Lozano*[a]

[a] *Department of Radiation Oncology, Institut Català d'Oncologia, L'Hospitalet, Barcelona, Spain*
[b] *Department of Hematology, Institut Català d'Oncologia, L'Hospitalet, Barcelona, Spain*
[c] *Department of Dermatology, Hospital Universitari de Bellvitge, L'Hospitalet, Barcelona, Spain*

ARTICLE INFO

*Article history:*
Received 26 March 2010
Accepted 4 May 2010

*Keywords:*
Langerhans cell sarcoma
Radiotherapy

ABSTRACT

We present the case of a patient with progressive Langerhans cell sarcoma whose cutaneous lesions and nodal masses were treated with palliative radiotherapy. Response to relatively low doses of radiotherapy was both good and sustained. We recommend a dose of 15–30 Gy depending on treatment intention and volume of the lesions.



## 1. Case report

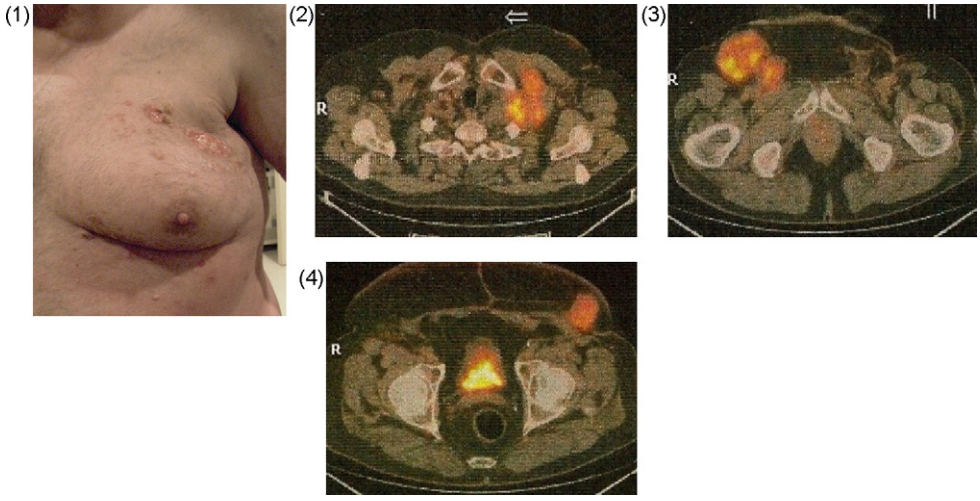
We present the case of a 63-year-old immunosuppressed patient diagnosed with Langerhans cell sarcoma in 2004. Relevant prior medical history includes a liver transplantation, for which the patient received immunosuppressive therapy. This current case report is a follow-up to a previously published report which described in detail the patient's indolent cutaneous lesions and nodal involvement.[1] From 2004 through 2007, the patient underwent multiple lymphadenectomies and surgical interventions to resect the skin nodules and diseased nodes. Histological analysis of various samples collected during this period confirmed the same diagnosis.

In 2007, the patient suffered a relapse, with disease progression evident at several nodal sites. As a result, weekly vinblastine chemotherapy was prescribed. Despite a partial response, the disease began to advance again 5 months after treatment. Chemotherapy was restarted, this time with a new regimen (CHOP scheme for 6 cycles). Despite a positive initial response, progression occurred again, this time after only 3 months after chemotherapy had been completed. Given the increasingly poor response to chemotherapy, the patient was referred to our radiation oncology department for possible palliative irradiation.

A physical examination showed bulky nodal masses located in the left supraclavicular area and bilateral inguinal region, and cutaneous lesions in the left thoracic wall (Images 1–4). The patient presented multiple skin lesions with small subcutaneous nodules; the larger nodules were exophytic and ulcerated. Additionally, he also complained of pain in the supraclavicular area.

A search of the literature revealed no previous reports on the use of radiotherapy in this type of tumour. The patient had been informed of this fact, but nevertheless agreed to proceed with treatment in the hope of alleviating his symptoms. We decided to treat the supraclavicular mass and the chest wall to relieve the pain and shrink the uncomfortable ulcerated cutaneous lesions. Radiotherapy was given over the left supraclavicular mass with 6 MV X-rays, 2 oblique AP-PA fields

* *Corresponding author*. Tel.: +34 93 260 77 22; fax: +34 93 260 77 25.
E-mail address: alucas@iconcologia.net (A. Lucas).
1507-1367/$ – see front matter © 2010 Greater Poland Cancer Centre, Poland. Published by Elsevier Urban & Partner Sp. z.o.o. All rights reserved
doi:10.1016/j.rpor.2010.05.001



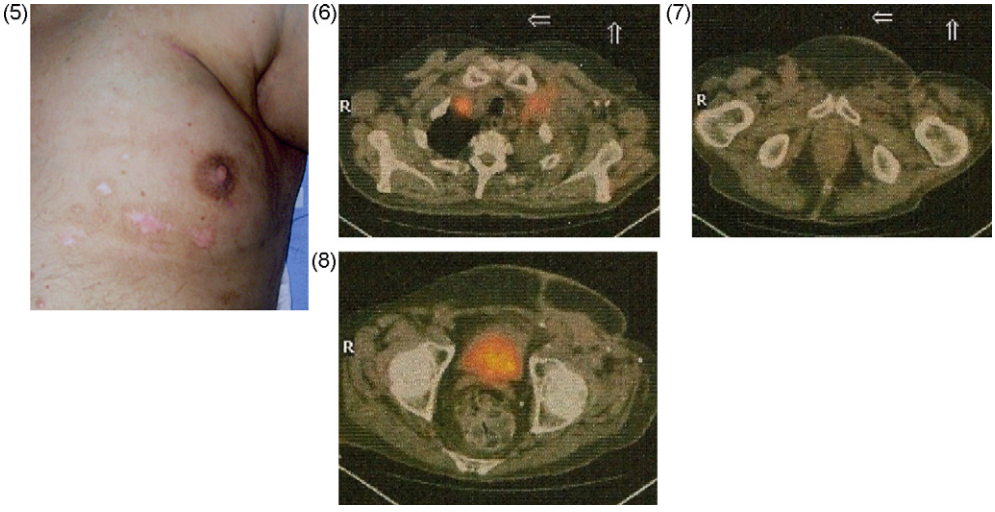

**Pictures 1–4 – Chest wall cutaneous lesions (1) and PET scan from supraclavicular (2) and inguinal (3 and 4) nodal masses before radiotherapy.**

of a Linear Accelerator Clinac 2100 (VARIAN). The left chest wall was treated with a 6 MeV direct electron field. In both cases, treatment was administered with a normofractionated scheme of 2 Gy per fraction, 5 days per week.

During the second week of treatment, it became evident that the lesions were responding well to treatment, with a response that continued to improve over the following days. After 36 Gy of radiotherapy had been delivered, the response of the mass and skin lesions was nearly complete. As a result, we decided that no further radiation was necessary and treatment was terminated. Tolerance was good, with the only side-effects being mild esophagitis and grade 2 dermatitis.

During the course of irradiation treatment, the patient complained of pain in the area of the bilateral inguinal masses. Given the good response achieved in the other locations, we decided to treat these areas as well. Two AP-PA fields of 18 MV X-rays were delivered over each site. The same dose was administered and at the end of treatment the response was also nearly complete. Side-effects included grade 2 dermatitis, grade 1 asthenia, and grade 1 anemia (shown on blood counts).

One month postradiotherapy, new cutaneous lesions appeared outside of the treatment field. A PET scan showed disease progression at several nodal sites and in the lung, pleura, and soft tissues. The patient was treated again with chemotherapy (cyclophosphamide, 50 mg/day and subcutaneous methotrexate, 43 mg), but he developed severe neutropenia and died of septic shock, 6 months after the first radiotherapy treatment (February 2009). At the time of death (August 2009), the irradiated areas of the skin continued to maintain the good response achieved by radiotherapy and, moreover, a PET scan showed a reduction in uptake in the supraclavicular area and absence of metabolic activity at inguinal regions (Images 5–8).




**Pictures 5–8 – Chest wall cutaneous lesions (5) and PET scan from supraclavicular (6) and inguinal (7 and 8) nodal masses after radiotherapy.**

## 2. Discussion

Langerhans cell sarcoma (LCS) is a rare malignant proliferation of Langerhans cells. LCS was first defined by the International Lymphoma Study Group in 2002.[2] Prior to that time, multiple terms were used to describe this entity, resulting in confusion. Since 2002, only a few cases have been described in the medical literature. Although those reports contain useful information about the histological features and clinical behavior of this neoplasm, information about treatment options is quite limited, as is often the case with rare diseases.

Histologically, cellular atypia and mitoses are needed to differentiate this disease from other Langerhans cell proliferations. Diagnosis also requires CD1a positivity and/or the presence of Birbeck granules.[3] Lee et al.[4] reviewed previous reports in 2006 and found only 19 cases described in English-language literature. Since that time, 6 new cases have been reported[1,5–10] (one of which was our patient, reported in 2007). Interestingly, a second case of a LCS patient with a transplanted liver was published in 2008.[7]

It appears that LCS occurs in a wide age range, though mainly in the third and fourth decades of life. Although the most commonly affected organs are the lymph nodes and skin, multiorgan involvement – which occurred in the case we present here – is also known to be characteristic. The lung, bone marrow, and spleen are the organs most often involved.

From published reports, we knew that LCS had an aggressive behavior in most cases. Unfortunately, information about the effectiveness of treatments is lacking. Surgery seems to be a good option for isolated lesions or confined nodal disease, but the benefit of adjuvant treatment is unknown. Chemotherapy has been used for the treatment of metastatic disease. Classical hematological schemes, such as CHOP and ESHAP, have been administered,[7] and other drug combinations with Adriamycin plus Ifosfamide, such as MAID (mesna, doxorubicin, ifosfamide, dacarbazine) have also been tested.[9,11,12] Nevertheless, the best option for systemic treatment of LCS has not yet been established.

Although we found no reports in the literature regarding the response of LCS to radiotherapy, we do know that Langerhans cell histiocytosis, the benign counterpart of LCS, responds well to low-dose radiotherapy. For this reason, we thought that radiotherapy might be useful as a local treatment, at least for pain relief and symptom control.

In the case we present here, the lesions began shrinking when the total dose reached 12–14 Gy, and response was almost complete when the total dose passed 30 Gy. At 36 Gy, a complete response was achieved and maintained for a long period. While lower doses may have been sufficient, we were encouraged to continue irradiating based on the low toxicity and good response to increasing doses.

As a result of our experience, we recommend this dose range in the treatment of these types of tumours, depending on treatment intention, the presence or not of gross tumour volume and its size.

## 3. Conclusions

LCS responds well to radiotherapy and we believe irradiation is a good option in selected cases with symptomatic or bulky lesions. Based on our experience, we suggest a dose range of 15–30 Gy, depending on the treatment intention and the volume of the lesions.

## Acknowledgments

The authors wish to thank Bradley Londres, at the Institut Català d'Oncologia, and Montse González for their assistance in editing this article.

## REFERENCES

1. Díaz-Sarrio C, Salvatella-Danés N, Castro-Forns M, Nadal A. Langerhans cell sarcoma in a patient who underwent transplantation. *J Eur Acad Dermatol Venereol* 2007;**21**(7):973–6.
2. Pileri SA, Grogan TM, Harris NL, Banks P, Campo E, Chan JK, et al. Tumours of histiocytes and accessory dendritic cells: an immunohistochemical approach to classification from the International Lymphoma Study Group based on 61 cases. *Histopathology* 2002;**41**(1):1–29.
3. Kawase T, Hamazaki M, Ogura M, Kawase Y, Maruyama T, Mori Y, et al. CD56/NCAM-positive Langerhans cell sarcoma: a clinicopathologic study of 4 cases. *Int J Hematol* 2005;**81**(4):323–9.
4. Lee JS, Ko GH, Kim HC, Jang IS, Jeon KN, Lee JH, et al. Langerhans cell sarcoma arising from Langerhans cell histiocytosis: a case report. *J Korean Med Sci* 2006;**21**(3):577–80.
5. Bohn OL, Ruiz-Argüelles G, Navarro L, Saldivar J, Sanchez-Sosa S. Cutaneous Langerhans cell sarcoma: a case report and review of the literature. *Int J Hematol* 2007;**85**(2):116–20.
6. Sumida K, Yoshidomi Y, Koga H, Kuwahara N, Matsuishi E, Karubek, et al. Leukemic transformation of Langerhans cell sarcoma. *Int J Hematol* 2008;**87**(5):527–31.
7. Yoshimi A, Kumano K, Motokura T, Takazawa Y, Oota S, Chiba S, et al. ESHAP therapy effective in a patient with Langerhans cell sarcoma. *Int J Hematol* 2008;**87**(5):532–7.
8. Deng A, Lee W, Pfau R, Harrington A, Di Giovani J, Prickett KA, et al. Primary cutaneous Langerhans cell sarcoma without Birbeck granules: indeterminate cell sarcoma? *J Cutan Pathol* 2008;**35**(9):849–54.
9. Uchida K, Kobayashi S, Inukai T, Noriki S, Imamura Y, Nakajima H, et al. Langerhans cell sarcoma emanating from the upper arm skin: successful treatment by MAID regimen. *J Orthop Sci* 2008;**13**:89–93.
10. Zhao G, Luo M, Wu YZ, Liu Q, Zhang B, Gao RL, et al. Langerhans cell sarcoma involving gallbaldder and peritoneal lymph nodes. *Int J Surg Pathol* 2009;**17**:347–53.
11. Ferringer T, Banks PM, Metcalf JS. Langerhans cell sarcoma. *Am J Dermatopathol* 2006;**28**(1):36–9.
12. Ohara G, Funayama Y, Satoh H, Uchida K. Chemotherapy for Langerhans cell sarcoma. *J Orthop Sci* 2009;**14**:242–3.