

Mantle Cell Lymphoma Relapsing at the Lymphedematous Arm

G. Massini¹, S. Hohaus¹, F. D'Alò¹, V. Bozzoli¹, B. Vannata¹, L. M. Larocca², L. Teofili¹

¹ Departments of Hematology, Catholic University, Rom, Italien; ² Departments of Pathology, Catholic University, Rom, Italy

▲ Introduction

Lymphedema (LE) is a chronic medical condition characterized by lymphatic fluid retention, resulting in tissue swelling. Cancer treatments involving lymph nodes can damage lymph drainage routes, causing accumulation of lymph fluid in the interstitial tissue of related limbs and body areas and secondary LE.

Of long term survivors (most patients undergone mastectomy), 0.07-0.04% can develop the Stewart-Treves syndrome, a rare and aggressive multifocal lymphangiosarcoma arising within the LE region, with a very poor prognosis. There are few cases reported in the literature on malignant lymphomas arising in a lymphoedematous arm, which can sometimes simulate the Stewart-Treves syndrome.

Case report

We report the case of a 45-year-old woman with stage IV mantle cell lymphoma (MCL) who was admitted to our hematology department. She had a massive LE of the left arm, as a consequence of previous breast cancer, treated four years before with radical mastectomy, radiation and chemotherapy. As lymphoma's therapy, she received four courses of R-CHOP, followed by three courses of R-MiCMA with haematopoietic stem cells (HSC) harvesting. The patient achieved complete remission and underwent autologous HSC transplantation as consolidation therapy. One year after HSC transplantation, the patient was again admitted to our department with a massive dissemination of purplish cutaneous nodules, in part ulcerated and infected, over the entire surface of the lymphedema-

Report on: Mantle cell lymphoma relapsing at the lymphedematous arm. *Mediterr J Hematol Infect Dis* 2013;5(1):e2013016.

tous arm (Fig.1), appeared on her left wrist two months before, and progressively sprouted over the entire arm without any systemic symptoms. The lesions were strictly confined to the area

of chronic LE, reminding the clinical picture of the Stewart-Treves syndrome (Fig.1). A total body CT scan was negative for nodal or parenchymal lymphoma localizations, and bone marrow biopsy showed no lymphoma cell infiltration. The skin biopsy showed multinodular infiltration of lymphomatous cells (Fig.1) that at immunohistochemistry resulted CD20/CD5/CyclinD1 positive, with a proliferative index of 90%. Patient received few courses of bortezomib and liposomal doxorubicin, without benefits; she died after few months with widespread pulmonary infiltration by MCL.

Discussion

The MCL is characterized by the striking tendency to disseminate throughout the body, infiltrating the lymphoid tissues, bone marrow and extranodal sites, so the aggressive clinical course observed in our patient is not surprising. Anyway, at relapse our patient had no localizations further than those in the lymphedematous arm. Hence, the awareness of the Stewart-Treves syndrome might help in the differential diagnosis of a suspicious cutaneous lesion within the LE.

In this case, we can presume that LE could have favoured the persistence of an occult localization of lymphoma, despite routine investigations evidenced an apparent complete remission of disease. Actually, the impaired lymphatic drainage could have made the involved area a "pharmacologic sanctuary", allowing lymphoma cells to skip the antiproliferative effect of chemotherapy. In addition, the eventual local immuno-deficiency, according to the hypothesis of the "immunocompromised district", might have promoted the chemotherapy resistance of malignant cells.

In conclusion, our experience shows that, in assessing the response to chemotherapy of lymphoma patient with chronic LE, it should be performed a careful investigation of the involved extremity to exclude residual tumour localizations.



Figure 1. The photo shows the massive dissemination of cutaneous lesions over the entire arm surface, appearing as confluent nodules, partially ulcerated and infected. All lesions were confined to the region of LE. The biopsy revealed the multinodular infiltration of lymphoid cells (hematoxylin and eosin staining, 40x magnification).

Correspondence Address

Giuseppina Massini, MD

Department of Hematology

Catholic University of Rome

Largo Gemelli 8

I-00168 Rom, Italy

E-Mail: giuseppina.massini@rm.unicatt.it