

Full Length Research Paper

An unusual presentation of non-Hodgkin lymphoma

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A 49 year old woman was admitted to our hospital with symptoms suggestive of breast cancer. The histological study of the left lumpectomy showed diffuse large B-cell lymphoma of the breast. In addition, the radiological assessment of the disease showed multiple organ involvement by the lymphoma. The patient achieved complete response after 3 cycles of chemotherapy but rapidly developed fatal evolution after the dissemination of the disease up to the central nervous system. This case confirmed the importance of the central nervous system prophylaxis in the situation of multiple extra nodal involvement of the diffuse large B-cell lymphoma.

Key words: Lymphoma, extra-nodal involvement, central nervous system involvement, central nervous system prophylaxis

INTRODUCTION

Lymphomas are malignant neoplasms of the lymphocyte cell lines. They mainly involve lymph nodes, spleen and other non-haemopoietic tissues. They are mainly classified as either Hodgkin's or non-Hodgkin's lymphoma (NHL) and of either B-lymphocyte or T-lymphocyte origin.

Disease of central nervous system (CNS) caused by the dissemination of haematological malignancies is an almost fatal complication (van Besien et al., 1998). The risk of CNS relapse ranged between 5 and 30% on all NHL subtypes.

The multiple organ involvement by the lymphoma is considered as a sign of high risk factor of CNS affection by the disease.

In this research, a case of unusual presentation (the disease was localized, at its first stage, in the breast, the thyroid gland, the kidney and the ovary) of non-Hodgkin lymphoma (NHL) with fatal evolution of the lymphoma after dissemination up to CNS was reported and the role of prophylaxis in the management of high grade NHL is

discussed.

CASE PRESENTATION

A 49 year old women (gravidia 0), at the state of menopause since at least one year, was admitted to our hospital with increasing lump in the left breast (having a 4 cm diameter at the first diagnosis). The patient, two months before diagnosis, had no inflammatory signs, no B symptoms (Fever, shudder, sweat and/or weigh loss) and had an ECOG performance status equal to 0.0 (Oken et al., 1982). The physical examination revealed a hard tumour measuring 4.8 cm in diameter. The tumour was located in the junction of the upper quadrants of the left breast. The enlarged left axillary lymph node measured 3 cm in diameter and the left thyroid nodule measured 4 cm in diameter. The mammography and ultrasonography are not suggestive of breast cancer: the first showed dense mass without micro-calcifications and the second showed heterogeneous well defined mass. In concordance with these results, the histological and immunohistochemical studies of the left lumpectomy showed diffuse large B-cell lymphoma (DLBCL) of the

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Figure 1a. CT scan of the neck showed thyroid involvement by malignant lymphoma.

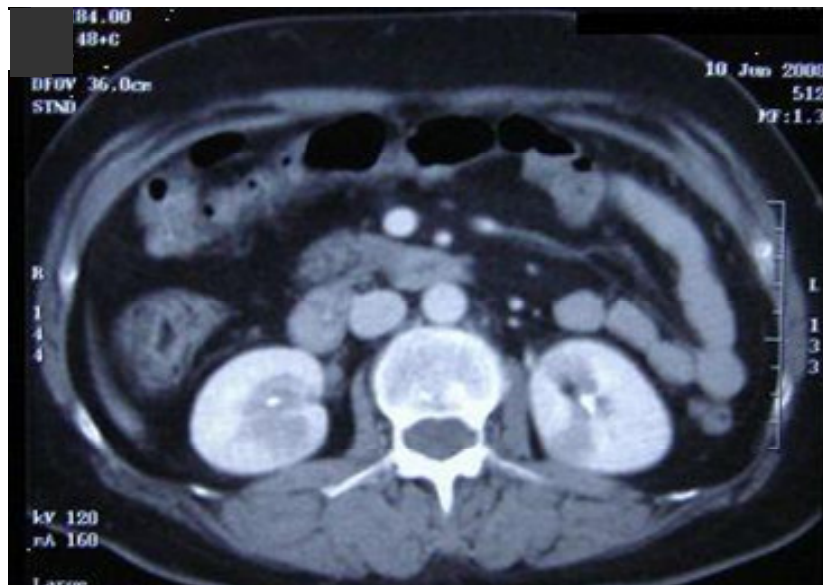


Figure 1b. CT scan of the abdomen showed left and right kidneys localisations.

breast following the Revised European-American Classification of Lymphoid Neoplasms/World Health Organisation classification of lymphoid neoplasms (REAL/WHO). Most of the neoplastic cells were positive for anti-CD-20 antibody. Computed tomography (CT) scan of the neck, chest, abdomen and pelvis in the postoperative period revealed multiple extra-nodal locations such as thyroid gland, ovary, spleen and kidneys (Figures 1a, 1b and 1c). The bone marrow biopsy showed no abnormalities. LDH (Lactate dehydrogenase) level was 535 U/L. The erythrocyte sedimentation rate was 28 mm in the first and 60 mm

in the second hour. The patient was staged IVAb according to the Ann Arbor staging system. Eighth cycles of CHOP (cyclophosphamide 750 mg/m² d1, doxorubicine 50 mg/m² d1, vincristine 1.4 mg/m² d1 and prednisone 50 mg/m² d1-5) treatment was planned. After 3 cycles of chemotherapy, the patient achieved a complete clinical and radiological response (observed on CT scan of the lung, abdomen and pelvis). The patient developed flask paraplegia after the fourth cycle of the chemotherapy. The magnetic resonance imaging (MRI) of the spin showed leptomeningeal infiltration of medullary cone



Figure 1c. CT scan of the pelvis showed right para-uterine mass suggestive of ovarian involvement by lymphoma.



Figure 1d. MRI showed infiltration of the leptomeninges by the lymphomatous process at the medullary cone and horse's tail

spinal decompression by laminectomy and tumour removal, the motor function did not recover. The histological study and immunohistochemistry showed large B-cell

non-Hodgkin's lymphoma of leptomeninges. The patient developed rapidly symptoms suggestive of lymphomatous meningitis and succumbed to disease 3 weeks after

the laminectomy.

DISCUSSION

The objective of this case presentation which was considered unusual DLBCL because of different localization (breast, thyroid, kidney, and ovary) of the lymphoma at the first manifestation of the disease was to increase the awareness of the oncologist." We can then make the right management with optimal prophylaxis against the risk of the fatal affection of CNS by the lymphoma (5 months after diagnosis in the present case). In fact CNS dissemination is an almost fatal complication of haematological malignancies. The risk of CNS relapse range between 5 and 30% on all NHL subtypes (Hollender et al., 2002). The median interval at relapse was between 3 and 9 months (interval range between 0 to 44 months). Given the high morbidity and mortality associated with CNS dissemination of NHL, a prophylactic strategy protocol is widely used against the CNS recurrence. This protocol comprises intrathecal methotrexate and systemic high dose methotrexate (up to 3 g/m²) in addition to conventional chemotherapy (CHOP based chemotherapy) (Hill et al., 2006; Hollender et al., 2002). In case of highly aggressive lymphoma (Burkitt lymphoma and lymphoblastic lymphoma), the prophylactic strategy is systematically indicated because the risk of CNS recurrence is higher up to 20% (Hill et al., 2006; Hollender et al., 2002 ; van Besien et al., 1998). In the case of DLBCL, the incidence of CNS recurrence in NHL is not sufficiently high (equal to 5%) to warrant the use of CNS prophylaxis in all patients (van Besien et al., 1998). However, the disease when in advanced stages induces the apparition of more than one extra nodal site and increases LDH serum level: The disease is then associated with higher risk of CNS recurrence with the probability of 25% (Hollender et al., 2002). The present case highlights the fatality of evolution of central nervous lymphoma and suggests the role of central nervous prevention in the case of DLBCL with multiple organ involvement and increase of serum LDH. We explain the no use of MTX to treat our patient by the lack of expertise in this field.

Conclusion

The involvement of more than one extra nodal site and the increase of serum LDH are associated with increased risk for CNS recurrence in NHL. Our case confirmed the indication of CNS prophylaxis in the situation of multiple extra nodal involvement of the DLBCL. The awareness of oncologists should be more acute in front of diffused large B cells lymphoma with high risk of dissemination.

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