

Concomitant nodal involvement by Langerhans Cell Histiocytosis and Hodgkin Lymphoma

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Introduction

Langerhans cell histiocytosis (LCH) is a clonal neoplastic proliferation of myeloid dendritic cells that upon activation migrate from the mucosal to lymph nodes. LCH is rarely, yet not exceptionally, found coexisting with other malignant neoplasms, suggesting it might arise in reaction to the cytokinic secretion of malignant cells.

Case Report

A 10-year-old girl presented with a right laterocervical mass that had been progressing for two months. Anamnesis revealed a weight loss of approximately ten percent over the last few months, mild permanent diffuse sweating and vague chest pains. Laboratory studies revealed an inflammatory anemia associated with elevated ESR and LDH.

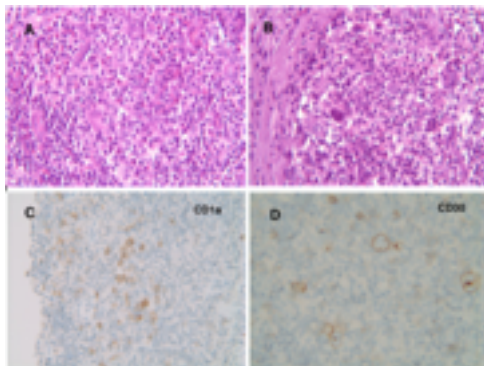


Fig 1. Cervical lymphadenopathy biopsy revealed a fibronodular pattern of eosinophilic inflammatory cells surrounding a few Reed-Steenberg cells CD30+ associated with numerous Langerhans cells and histiocytes CD68+.

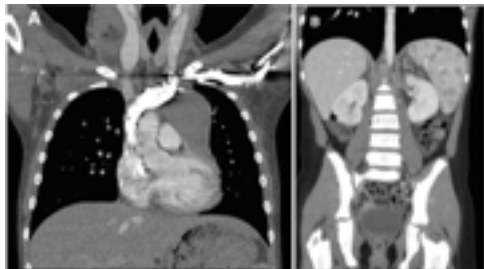


Fig. 2. Initial imaging by computed tomography showed bilateral jugular lymphadenopathy, a large mediastinal mass, multiples splenic nodules, and numerous mediastinal and lomboarctic lymphadenopathies.

These elements pleaded for the diagnosis of scleronodular Hodgkin Lymphoma associated with LCH. The patient was treated according to the Euronet PHL C1 TL3 protocol. Response evaluation by PET-CT after six cycles of chemotherapy revealed complete remission.

Discussion

Coexistence of neoplasms is an exceptional phenomenon, yet already described with LCH [1].



Some studies suggest a 0.3% rate of occurrence of the association, whether meta- or synchronous, of LCH with Hodgkin lymphoma, which can be considered significant regarding the low incidence of both diseases. This suggests that the two phenomena are related. The current prevalent hypothesis is to consider LCH a reactive process to neoplasms, particularly when associated with Hodgkin lymphoma, since both disease involves the same node concomitantly. This is also supported by the primary regression of LCH during treatment and the finding of polyclonal Langerhans cells when associated with malignant neoplasms [2]. The proliferation of Langerhans cells could be linked to cytokines secreted by the inflammatory background cells of Hodgkin lymphoma.

Take Home Messages

- Various neoplasms have been reported associated with LCH. Timing of occurrence is different for LCH associated with lymphoma and leukemia, suggesting distinct mechanisms underlying these associations.
- Practicians should be aware of the possibility of the combination of LCH with Hodgkin lymphoma, since both diseases can take the same clinical form, and yet have a very different prognosis.
- Diagnosis of isolated LCH should be posed after exclusion of underlying more aggressive diseases.

References:

- [1] Egeler RM, Neglia JP, Puccetti DM, et al. Association of Langerhans Cell Histiocytosis with Malignant Neoplasms. *Cancer* 1993;71:865-73.
- [2] Christie LJ, Evans AT, Bray SE, et al. Lesions resembling Langerhans cell histiocytosis in association with other lymphoproliferative disorders: a reactive or neoplastic phenomenon? *Human Pathology* (2006) 37, 32-39