Crystal-Storing Histiocytosis in a Patient with Nodal Marginal Zone Lymphoma

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Abstract
We present a case of a 72-year-old male diagnosed with concurrent crystal-storing histiocytosis and nodal marginal zone lymphoma with plasmacytic differentiation following a finding of new axillary lymphadenopathy during active surveillance for an early stage lung cancer. A panel of ancillary studies was performed to help establish the diagnosis and to exclude other histologic mimics. We share this case to promote awareness of this rare entity, discuss its association with immunoglobulin-producing lymphomas and review an approach to cases with CSH to avoid misdiagnosis.

Keywords
Crystal-storing histiocytosis, Nodal marginal zone lymphoma, Plasmocytic differentiation

Abbreviations
CSH: Crystal-Storing Histiocytosis; NMZL: Nodal Marginal Zone Lymphoma; CT: Computed Tomography; PAS: Periodic Acid-Schiff

Introduction
Crystal-storing histiocytosis (CSH) is a rare disorder in which crystallized material accumulates within the cytoplasm of histiocytes. Many authors have proposed that the crystallized material represents an intralysosomal accumulation of immunoglobulins [1,2], as most cases of CSH are associated with B-cell lymphomas with plasmacytic differentiation or plasma cell neoplasms [3-5]. In some cases, a dominant histiocytic component can obscure the underlying neoplasm, which can possibly lead to an error in diagnosis. Herein, we present a case of nodal marginal zone lymphoma (NMZL) with plasmacytic differentiation occurring in association with CSH in a 72-year-old male with incidental axillary lymphadenopathy. In this article, we also discuss our approach to ancillary testing in cases of CSH, which is essential for excluding a variety of its histologic mimics.

Case Report
A 72-year-old male status post radical radiotherapy for an early stage lung cancer was found to have new axillary lymphadenopathy on a computed tomography (CT) scan during active surveillance. Serum electrophoresis showed no paraprotein. Serum-free light chains were within normal limits. A positron emission tomography scan showed no FDG avid bone marrow or splenic involvement.

A fine needle aspiration of a right axillary lymph node revealed a cellular sample showing a mixture of small and medium sized lymphoid cells, scattered plasmacytoid cells, eosinophils, and plenty of large polygonal shaped histiocytes with abundant finely granular cytoplasm. Immunophenotypic analysis by flow cytometry confirmed a monotypic (lambda restricted) mature B-cell neoplasm, which were CD5 and CD10 negative.

Histopathologic examination of the excised lymph node showed an effaced architecture with small sized atypical lymphocytic infiltrates, foci of microgranulomas and sheets of histiocytes with abundant granular cytoplasm.

By immunohistochemistry, the small lymphoid tumour cells were positive for CD20, but lacked staining for CD5 and CD10 (Figure 1). Scattered plasma cells...
CD1a, CD30, SOX10, desmin, Factor XIIIa, and EBER-ISH were negative (Figure 3). PCR showed a negative result for a MYD L265P mutation.

A diagnosis of a NMZL with plasmocytic differentiation were also identified, which were monotypic for lambda light chains. The histiocytes were positive for CD68. PAS, PASD, and AFB were negative for storage disease and mycobacteria, respectively (Figure 2). Cytokeratin, S100, CD1a, CD30, SOX10, desmin, Factor XIIIa, and EBER-ISH were negative (Figure 3). PCR showed a negative result for a MYD L265P mutation.

A diagnosis of a NMZL with plasmocytic differentiation...
Figure 3: Crystal-storing histiocytosis showing positivity for CD68 (A, 100X). Special stains, including AFB (B, 100X), PAS (C, 100X) and PASD (D, 100X), were negative.

Figure 4: Sheets of histiocytes with granular cytoplasm are negative for CD1a (A, 100X), S100 (B, 100X), factor XIIIa (C, 100X), SOX10 (D, 100X) and desmin (E, 100X).
We also emphasize the observation that tissues with extensive histiocytosis may obscure an underlying lymphoid neoplasm. An awareness of this association is therefore crucial to avoid such misdiagnosis. The present case adds to the few reports in the literature of CSH associated with a NMZL and highlights the importance of having a high degree of awareness of this rare entity.

Disclaimers

Authors declare that there are no conflicts of interest regarding the publication of this paper.

References