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TEACHING CASES

Primary splenic marginal zone lymphoma with florid granulomatous reaction—A case report and review of literature

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Abstract

Splenic marginal zone lymphomas (SMZL) constitute about 20% of primary splenic NHLs. We report a case of primary SMZL with a florid granulomatous reaction which obscured the underlying lymphoma. Although granulomas have been described in splenic non-Hodgkin lymphoma, it can be extensive and mask the underlying lymphoma. A careful search for the cytoarchitectural features of SMZL is warranted in such a case.

Keywords: Splenic marginal zone lymphoma; Granuloma

Introduction

Splenic marginal zone lymphoma (SMZL) constitutes less than 1% of all non-Hodgkin lymphomas (NHLs) and about 20% of primary splenic NHLs [4]. We report a case of primary splenic NHL with histologic and immunophenotypic features of primary SMZL with a florid granulomatous reaction in the spleen and involved splenic hilar nodes. Although a granulomatous reaction has been described in NHL of the spleen, only rarely does it acquire such proportions as to obscure an underlying lymphoma.

Clinical history

A 45-year-old man was diagnosed to have autoimmune hemolytic anemia in December 2002 and was on steroids and later on azathioprine. Subsequently, he developed azathioprine-induced skin rashes, for which his therapy was discontinued, and he was started on mycophenolate. Although his hemoglobin concentration had increased with these drugs, he had developed progressively increasing spleen during this period. In March 2006, he was admitted for a therapeutic splenectomy. At the time of admission, he was afebrile and icteric. There was no peripheral or intraabdominal lymphadenopathy. He was found to have mild hepatomegaly of 1.5 cm and massive splenomegaly of 20 cm in size. In March 2006, his hemogram was as follows: hemoglobin – 11.2 g%, platelets – 145×10^9/L, total WBC count – 19.1×10^9/L with 48% atypical lymphoid cells. Direct Coomb's test was positive. Peripheral blood immunophenotyping results were as follows: CD19 – 81.5%, HLA DR – 86.1%, SmIg – 87.4%, lambda – 84.6%, CD23, CD10- and CD5-negative, which was consistent with a B-cell lymphoproliferative disorder. His lupus anticoagulant was positive at presentation. There was no reversal of serum albumin/globulin (A/G) ratio. His chest X-ray was normal. Abdominal

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ultrasonography showed massive splenomegaly with enlarged splenic vein without any significant lymphadenopathy. There was no clinical or radiological evidence of tuberculosis or sarcoidosis. He was HCV-negative. Therapeutic splenectomy was done.

Materials and methods

The specimen received was a massively enlarged spleen (2250 g). The external surface was unremarkable. Sectioning revealed multiple small gray white nodules ranging from 0.1 to 0.5 cm in diameter. One hilar lymph node was identified, 1.5 cm in diameter, with a gray–white soft cut surface. Histologic sections were prepared from 10% formalin-fixed, paraffin-embedded tissue and stained with hematoxylin and eosin. Immunohistochemical stains were performed using the avidin-biotin technique (see Table 1 for the list of antibodies used).

Results

Hematoxylin and eosin-stained sections (4 μ) from the spleen showed a micronodular pattern with expansion of white pulp with enlarged splenic follicles, some of which were coalescent (Fig. 1). The follicles were composed of small lymphoid cells with clumped chromatin towards the center and medium-sized cells with round nuclei and visible nucleoli and moderate amounts of clear cytoplasm towards the periphery of the follicle (Fig. 2). Some of the white pulp nodules showed residual germinal centers with hyaline, amorphous deposits. The infiltrate spilled into the red pulp with much cordal lymphocytosis, with cytologic features similar to the small lymphoid cells occupying the white pulp centers. There were numerous discrete and confluent well-defined granulomata composed of epithelioid histiocytes and foreign body and Langhans' type giant cells centered on the white pulp tumor nodules and some around it. In areas, the confluent granulomata effaced the splenic architecture across contiguous low power fields (Fig. 3). The confluent granulomata were seen around trabeculae as well. There were foci of fibrinoid necrosis and hyalinization within these confluent granulomata. There was no evidence of caseous necrosis and large cell transformation. Although few scattered plasma cells were seen, significant plasmacytic differentiation was not present. Dutcher bodies were not identified. Special stains for acid-fast bacilli done on multiple sections of the spleen and the hilar lymph node were negative. Sections of the hilar lymph node showed

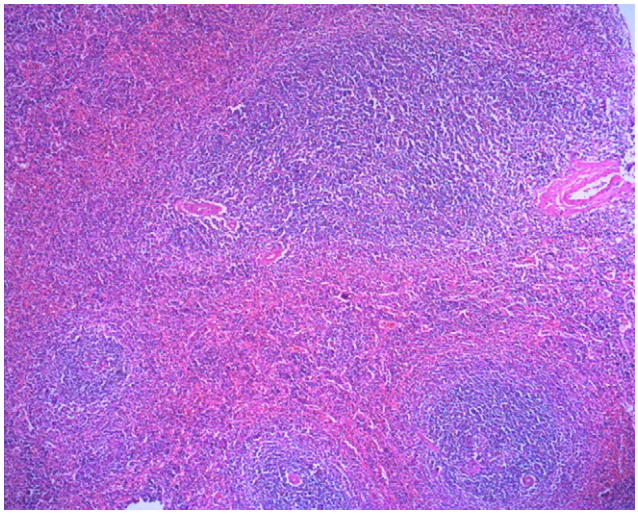

Fig. 1. H&E, X50. Spleen with enlarged white pulp and spillage of neoplastic lymphoid cells into the red pulp.

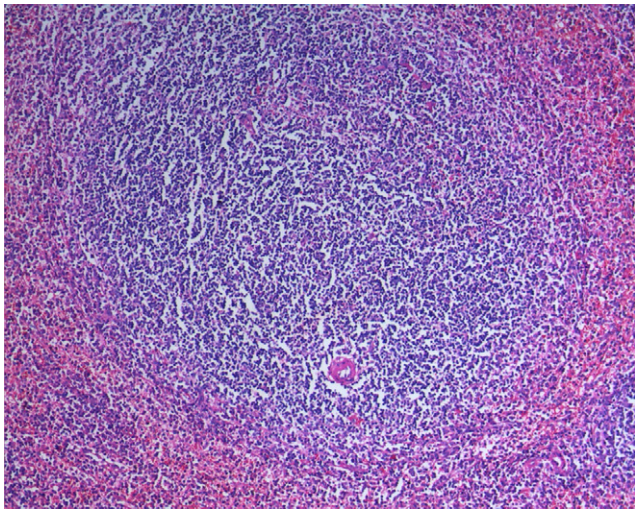

Fig. 2. H&E,X100. Neoplastic follicle with a population of small cells towards the center and medium-sized cells with clear cytoplasm at the periphery.

Table 1. Antibodies used for immunohistochemical studies

Name of antibody	Clone	Dilution	Source
CD20	Clone L26	1:400	Dakocytomation
CD3	Clone F7.2.38	1:75	Dakocytomation
Ki-67	Clone MIB-1	1:75	Dakocytomation
CD10	Clone SS2/36	1:80	Dakocytomation
CD5	Clone CD5/54/F6	1:50	Dakocytomation
CD23	CloneMHM6	1:10	Dakocytomation
CYCLIN D1	CloneDSC-6	1:200	Dakocytomation
Bcl-2	Clone124	1:80	Dakocytomation