

# Primary Splenic Hodgkin Lymphoma Presenting with Chronic Pruritus

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## Introduction

Hodgkin Lymphoma is an uncommon B-cell lymphoma which accounts for 11.5% of all lymphomas. The majority of patients present with lymphadenopathies, and only one-third of patients are referred with fever, night sweats, weight loss and chronic pruritus. The most common extranodal sites involved in Hodgkin lymphoma are spleen, lung, liver and bone marrow.<sup>1</sup> Spleen is the most frequent organ in abdomen which is involved in Hodgkin lymphoma,<sup>2</sup> and sometimes it may be the only abdominal organ involved.<sup>3</sup> But only splenic involvement without lymphadenopathies is very rare as the presenting feature of Hodgkin lymphoma. In a study of 184 cases of lymphoma in Japan, only 1.09% were involved by Hodgkin lymphoma.<sup>4</sup>

## Case Report

A 69-year-old man presented with chronic generalized pruritus from 6 months ago. Multiple topical and systemic treatments had been administered for skin as a source of itching, and the symptoms had subsided for 2 months. Then, night sweats, fever and shaking, loss of appetite, weight loss and vomiting were added to his symptoms and itching started again. The patient underwent work up for pancytopenia and weight loss. Physical examinations showed excoriations on the skin and only mild splenomegaly with no other lymphadenopathies. Complete blood count analysis showed: WBC: 1200/  $\mu$ L, RBC: 3.75/  $\mu$ L, Hb: 9.6 g/dL, Plt: 27000/  $\mu$ L. Upper gastrointestinal endoscopy was done and revealed erythema in the greater curvature mucosa of stomach with moderate to severe chronic gastritis and positive for *H. pylori* infection on biopsy. Abdominopelvic sonography displayed enlarged spleen, M: 179 mm in greatest diameter, mild fatty changes in liver and was otherwise unremarkable. Abdominal CT scan confirmed multiple hypodense nodules in spleen with no other thoracic or abdominal lymphadenopathy. Bone marrow aspiration and biopsy of the patient showed normocellular marrow with normal maturation of hematopoietic cells.

Due to refractory anemia, thrombocytopenia, normal bone mar-

row examination and upper GI endoscopy and persistent epistaxis, the patient was candidate for splenectomy.

## Pathological findings

Received specimen in formalin consisted of one spleen M: 16  $\times$  12  $\times$  4 cm, weight: 550 g. Cut sections showed multiple tiny white nodules in parenchyma (Figure 1).

Sections from paraffin-embedded blocks revealed multifocal discrete nodules in which there was a polymorphic population of hematopoietic cells, mainly composed of small mature looking lymphocytes admixed with few eosinophils, plasma cells and histiocytes as well as few scattered large bizarre cells resembling Hodgkin cells and Reed-Sternberg cells. Some mitotic figures were also seen (Figure 2)

immunohistochemical stainings were done on paraffin-embedded tissue samples of the spleen. The above mentioned cells were positive for CD30 and negative for CD20, CD3, LCA and CD15 (Figure 3).

Morphologic study and IHC staining were in keeping with classical Hodgkin lymphoma.

## Discussion

Involvement of the spleen is common in disseminated Hodgkin lymphoma and non-Hodgkin lymphoma; however, spleen is a rare primary site of involvement by Hodgkin's and Non-Hodgkin's Lymphomas.<sup>5-7</sup> Furthermore, chronic pruritus is even less common as the presenting symptom of Hodgkin lymphoma.<sup>1</sup> In this special case, we have two uncommon features together which consist of pruritus as the first presenting symptom and the only organ involvement in the spleen. In literature review, we found only few cases of primary splenic Hodgkin lymphomas although some of these cases had liver involvement, too.<sup>8-11</sup> In three cases with only splenic involvement, one was detected in a young man and in the settings of immune deficiency (chronic granulomatous disease), who also presented with fever and splenomegaly.<sup>12</sup> Another case was a 15-year-old boy with traumatic rupture of spleen and incidental finding of Hodgkin lymphoma in his spleen without any nodal involvement.<sup>13</sup> The latter had the same presentation as our case.<sup>14</sup> Regarding the documented cell-mediated immune response of spleen in alloantigens, the possible role of this organ in tumoral immunosurveillance should be also considered.<sup>15</sup> It seems reasonable to use sensitive methods such as PET scan for detecting splenic lesions suspicious of lymphomatous involvement, especially in elderly patients presenting with prolonged fever with no identifiable infectious origin.<sup>16</sup> Although the presence of fever

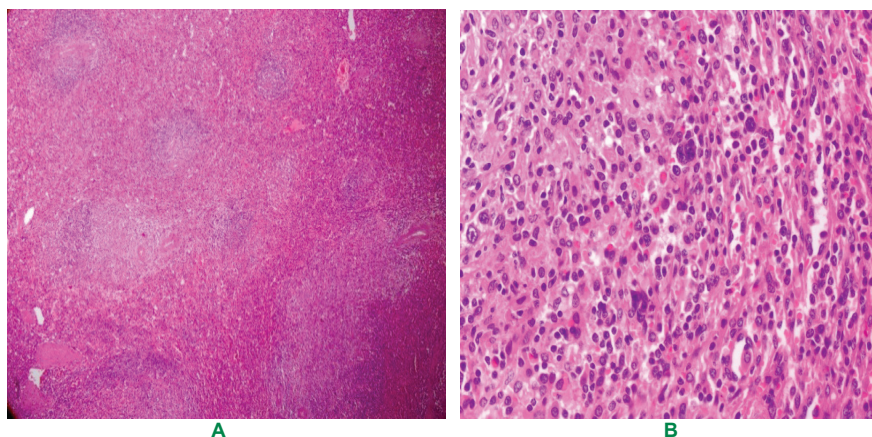
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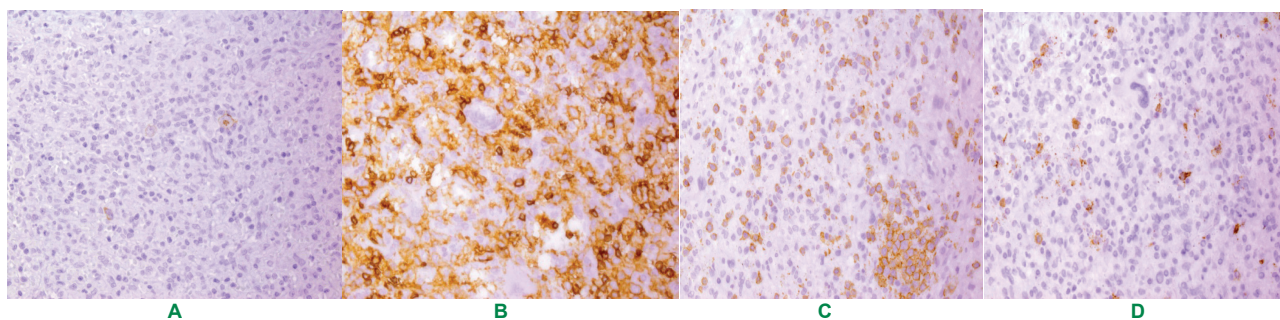
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**Figure 1.** Multiple tiny whitish discrete nodules in spleen parenchyma sectioning.



**Figure 2.** Sections of spleen show multifocal discrete nodules in parenchyma and remnants of white pulp (A. H&E  $\times 40$ ), Reed-Sternberg cells are seen in the background of mixed inflammatory cells (B. H&E  $\times 400$ )



**Figure 3.** Reed- Sternberg cells negative for CD3 (A), CD 20 (B) and LCA (D). Positive staining for CD 30 (C).

and miliary splenic nodules in most of these cases may be seen accidentally, this approach may help in earlier diagnosis of Hodgkin lymphomas. Since these patients have been symptom-free only after splenectomy or short courses following chemotherapy, early diagnosis could help in choosing the most appropriate treatment plan. No statistics are now available to compare their survival with other classic Hodgkin lymphomas with generalized nodal involvement. This hypothesis could open a new window to more precise evaluation of spleen in febrile old patients with constitutional symptoms and also in sectioning splenectomy specimens in pathobiology laboratories.

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