Case Report

Primary splenic lymphoma masquerading as splenic abscess: A rare case

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ABSTRACT

The incidence of primary splenic lymphoma (PSL) is <1%. Here we report a case of PSL in a 63-year-old female patient who had fever, night sweats, anorexia, and weight loss for 2 months. Abdominal contrast-enhanced computed tomography showed mild splenomegaly with multiple patchily enhancing splenic space occupying lesions (SOLs) along with multiple enlarged splenic hilar lymph nodes. A provisional diagnosis of splenic abscesses was made. Following splenectomy, histopathological examination and immunohistochemical analysis revealed this to be a case of splenic non-Hodgkin lymphoma – diffuse large B-cell – nongerminal center type. This case is significant since it was misdiagnosed as splenic abscess initially. Even though, PSL is an extremely rare tumor, this entity must be borne in mind when splenic SOL is detected radiologically. Documentation of such rare cases is of utmost importance to facilitate early diagnosis and treatment of similar cases in the future.

Key words: Abscess, Primary splenic lymphoma, splenic space occupying lesion

INTRODUCTION

Primary splenic lymphoma (PSL) is an extremely rare neoplasm with a reported incidence of <1%.[1] PSL is difficult to diagnose and the common differential diagnoses of the condition include hemangioma, lymphangioma, abscess, hamartoma, infarct and metastatic disease.[2] Here, we present a case of PSL in a 63-year-old female patient, which was initially diagnosed as splenic abscess.

CASE REPORT

A 63-year-old hypertensive female patient presented with fever, night sweats, anorexia and weight loss for a period of 2 months.

General examination revealed the presence of pallor. Abdominal examination showed that spleen was palpable for approximately 7 cm below the left costal margin. The surface of spleen was nodular, and its consistency was firm. No other significant finding was noted after thorough systemic examination.

Complete blood count revealed that the patient had microcytic hypochromic anemia (hemoglobin level – 8 g/dL). The total and differential counts were within normal limits. Liver function tests were unremarkable except for the slightly raised level of serum lactate dehydrogenase (140 U/L). Antinuclear antibodies and markers of hepatotropic viruses were absent.

Abdominal contrast-enhanced computed tomography (CECT) showed mild splenomegaly with multiple patchily enhancing splenic space occupying lesions (SOLs), of which the largest measured 6.4 cm × 4.8 cm. Multiple enlarged splenic hilar lymph nodes were also found with matting and central necrosis in a few nodes [Figure 1]. On the basis of radiologic features, a provisional diagnosis of splenic abscesses was made.

Splenectomy was performed along with excision of splenic hilar lymph nodes when the patient failed to respond to medical treatment for a period of 1½ months. Gross examination of the resected specimens showed that the spleen was approximately 10 cm × 8 cm × 5 cm in size. Cut

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section revealed multiple grayish white nodules, the largest one measuring 4.5 cm in diameter. Three lymph nodes were also sent, of which the largest measured approximately 3.5 cm in diameter [Figure 2a].

Microscopic examination of sections from spleen and lymph nodes showed nodular areas of various sizes surrounded by fibrosis. In focal areas, the nodules were found to encroach on the splenic capsule. The nodules were composed of predominantly large lymphoid cells with pleomorphic nuclei, centrally placed nucleoli, peripherally condensed chromatin and a moderate amount of amphophilic cytoplasm. Multinucleated giant cells were also present along with occasional lymphocytes and plasma cells. Few atypical mitotic figures were seen. Areas of necrosis were also noted [Figure 2b]. Based on the histopathological features, a diagnosis of splenic lymphoma (non-Hodgkin's lymphoma [NHL] – large cell type) was rendered.

Subsequently, immunohistochemical analysis was undertaken. CD3 was positive only in the reactive lymphocytes. The tumor cells were positive for CD20 and negative for Bcl-6. Ki-67 positivity was noted in 25–30% tumor cells [Figure 3]. Based on these findings, the tumor was diagnosed as splenic NHL – diffuse large B-cell – nongerminal center type.

Postoperatively, the patient received cyclophosphamide, adriamycin, vincristine, and prednisone (CHOP) chemotherapy. She has been on regular follow-up for the last 1 year, which was uneventful.

DISCUSSION

The definition of PSL remains a matter of controversy. The oldest definition was provided by Dasgupta *et al*. and they defined PSL as a condition confined to spleen or hilar lymph nodes with no recurrence of disease for at least 6 months after splenectomy.[3] In the present case, the patient fulfilled the criteria of this definition. A broad definition of PSL was proposed by Kehoe and Straus. And they included patients with simultaneous minimal involvement of liver and lymph nodes beyond the splenic hilum, within the canvas of PSL, so long as the bulk of the disease remained within the spleen.[4]

The etiology of PSL remains largely unknown. It has been suggested that chronic hepatitis C virus infection along with some poorly defined genetic and environmental factors play a significant role in its development.[5]

The common clinical features of PSL may be variable, some presenting with fever, weight loss, weakness and left upper quadrant discomfort, thereby mimicking splenic abscess.[6] Ingle and Ingle. recently reported a case of PSL in a middle aged female who presented with massive splenomegaly and hypersplenism. This prompted the surgeon to undertake therapeutic splenectomy.[7] In the present case, the patient presented with fever, night sweats, anorexia, weight loss and splenomegaly.

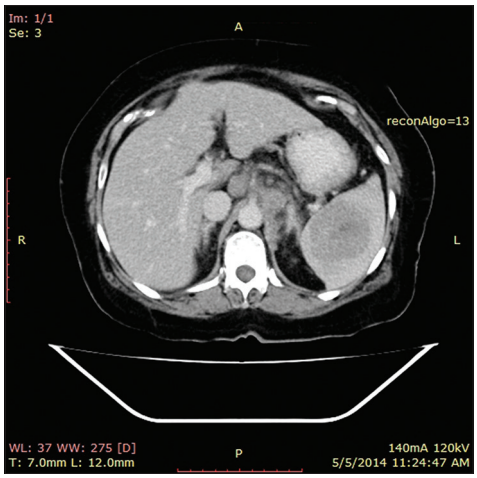

Figure 1: Computed tomography scan picture showing mild splenomegaly with multiple patchily enhancing splenic space occupying lesions

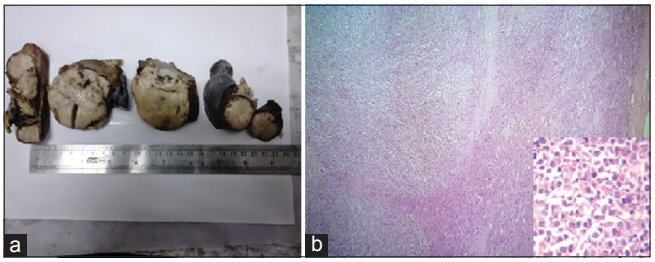

Figure 2: (a) Gross appearance of spleen showing multiple grayish white nodules, (b) primary splenic lymphoma-nodular areas of various sizes surrounded by fibrosis (H and E, ×100); inset shows large lymphoid cells of the tumor with pleomorphic nuclei, prominent nucleoli and peripherally condensed chromatin (H and E, ×400)

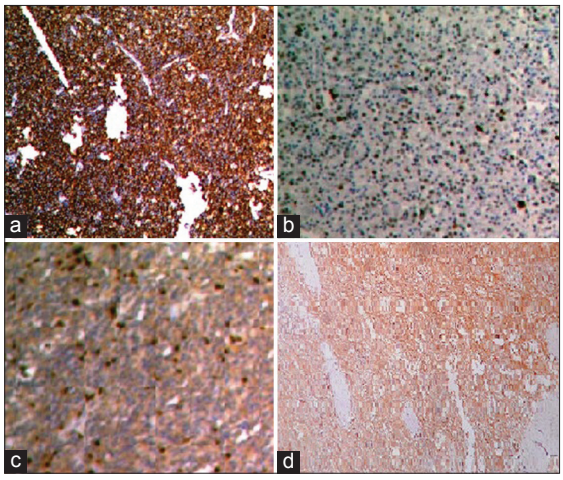

Figure 3: Primary splenic lymphoma (immunohistochemistry) – (a) CD20 positive tumor cells (×100), (b) Ki-67 positivity in 30% tumor cells (×100), (c) CD3 positive reactive lymphocytes (×100), (d) Bcl-6 negative tumor cells (×100)

The radiologic investigation of choice for the diagnosis of PSL is CECT. Most lesions appear hypoechoic, but sometimes anechoic areas are seen, which suggest liquefactive necrosis thereby causing confusion with splenic abscess.[2] In this case, CECT showed mild splenomegaly with multiple patchily enhancing splenic SOLs. On the basis of radiologic features, a diagnosis of splenic abscess was given.

The differential diagnoses of solitary splenic lesion include a large number of entities. Hemangioma, lymphangioma and hamartoma are the most frequently encountered diagnoses in case of a focal splenic lesion. If the patient is afebrile and symptomatic, then the most likely diagnosis is a metastatic lesion. In the case of an acute ischemic lesion, the condition is painful and clinical evidence of the predisposing cause is usually present. In febrile or immunocompromised patients, splenic abscess is a possible diagnosis. Since patients of PSL usually have no lymphadenopathy, and their peripheral blood pictures remain normal, diagnosis becomes difficult.[2]

The gross appearance of splenic lymphoma has been classified into four categories: (1) Homogeneous enlargement without masses, (2) miliary masses, (3) 2–10 cm masses and (4) large solitary mass. A staging system of PSL has also been proposed: Stage I – Tumor confined to spleen only; Stage II – Involvement of spleen and hilar lymph nodes; Stage III – Involvement of extra splenic nodes or liver.[8] In this case since the hilar lymph nodes were involved, the patient had Stage II disease.

It has been stated that all kinds of cells of malignant lymphoma can occur in PSL. The prognosis of small cell lymphoma has been found to be slightly better than intermediate-grade lymphoma.[6]

The present case was morphologically diagnosed as NHL – large cell type. A final diagnosis of diffuse large B-cell, nongerminal center type of NHL was rendered only after immunohistochemical analysis with CD3, CD20, Bcl-6 and Ki-67. Konstantiadou *et al*. Confirmed their diagnosis of anaplastic large B-cell type of NHL of spleen after obtaining positive immunohistochemical results with CD20 and CD79a.[2] Hans *et al*. Classified diffuse large B-cell lymphoma on the basis of expression of Bcl-6. Cases that were positive for Bcl-6 were termed germinal center type, and they were found to have a better prognosis in comparison to the Bcl-6 negative cases.[9] The present case was classified according to this scheme.

There are controversies regarding the best treatment protocol for PSL. The modalities used include Splenectomy only, splenectomy followed by chemotherapy, splenectomy followed by radiotherapy or a combination of chemotherapy and radiotherapy.[6] The most commonly used chemotherapeutic regimen is CHOP. Some authors are of the opinion that overall survival rates are better with early splenectomy, followed by multidrug chemotherapy compared with splenectomy alone or splenectomy, followed by single agent chemotherapy.[10]

The present case is interesting due to rarity of occurrence of PSL. Documentation of such rare cases along with details of treatment prescribed is the cornerstone for defining optimum diagnostic and therapeutic strategies.

CONCLUSION

Primary splenic lymphoma is an extremely rare entity. Radiological and clinical appearances of PSL can mimic splenic abscess that may delay the proper diagnosis and management. Hence, it is imperative that clinicians keep this differential diagnosis in their minds while dealing with similar cases and undertake necessary steps like biopsy and immunohistochemical analysis to reach the correct diagnosis.

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