

## PRIMARY LYMPHOMA OF THE SPLEEN. REPORT OF A CASE WITH DIAGNOSIS BY FINE-NEEDLE GUIDED BIOPSY

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### ABSTRACT

Primary splenic lymphoma is a relatively infrequent disease; the diagnosis of this entity is currently made with splenectomy. In a 52-year-old female with left upper quadrant abdominal pain, ultrasound showed a normal-sized spleen with an internal hypoechoic focal lesion. Ultrasonically-guided fine-needle aspiration and tissue core biopsy of the splenic lesion showed non-Hodgkin's lymphoma (NHL). At the time of presentation there was no evidence of involvement of lymph nodes, bone marrow or any other organ. A diagnosis of primary splenic non-Hodgkin's lymphoma was made and the patient underwent laparotomy with splenectomy. Histologic examination of the spleen confirmed the diagnosis: low-grade NHL confined to the spleen. The patient is well and in complete remission seven months after diagnosis.

The purpose of this paper is to report a rare occurrence of primary splenic lymphoma and to demonstrate the possibility of making this diagnosis by percutaneous guided biopsy.

Key words: primary splenic lymphoma, fine-needle biopsy

Malignant lymphoma seldom presents as a primary neoplasm of the spleen. The spleen is involved in approximately 25% of patients with lymphoma at initial staging;<sup>1</sup> otherwise, splenic involvement is common in advanced stages of Hodgkin's disease (HD) and non-Hodgkin's lymphoma (NHL).<sup>2</sup> Since secondary involvement of the spleen in lymphoma is quite common, the definition of primary splenic lymphoma (PSL) is still used ambiguously in the literature, as recently reported by Gobbi *et al.*<sup>3</sup>

Das Gupta *et al.*<sup>4</sup> have suggested strict criteria for establishing a diagnosis of PSL. They proposed that this diagnosis be made when the disease was confined to the spleen or hilar lymph nodes, and no recurrence of the disease is evident for at least 6 months after splenectomy.<sup>4</sup>

The case reported below not only met these strict criteria, but demonstrated the possibility of making the diagnosis by percutaneous, ultrasonically-guided biopsy.

### Case report

A 52-year-old female was admitted with a two-month history of left upper quadrant abdominal pain and night sweats.

She denied having a history of weight loss, fever or chills. Her physical examination was essentially unremarkable. Laboratory work-up, including lactate dehydrogenase, complete blood cell count, ESR, serum electrophoresis and immunoelectrophoresis, urinalysis, renal and liver function were within normal limits.

Ultrasound (US) examination of the abdomen

revealed a hypoechoic focal lesion in the spleen, 2.5 cm in diameter, while the spleen itself showed normal dimensions: 12×9×4 cm. No splenic-hilar or abdominal adenopathy was seen. A total body computerized axial tomography (CT) study confirmed the splenic lesion (Figure 1), without evidence of other lesions or thoracic and/or abdominal adenopathy. Ultrasonically-guided fine-needle aspiration biopsy was carried out on the splenic mass according to previously reported principles,<sup>5</sup> and revealed NHL (Figure 2).

Bone-marrow biopsy was negative; US-guided tissue core biopsy of the splenic lesion was performed,<sup>6</sup> and histologic examination confirmed low-grade type NHL. A diagnosis of PSL was made and the patient underwent splenectomy. The spleen presented normal dimensions and a 2.2 cm focal mass, as previously detected by US and CT examination. No abdominal adenopathies were found and two liver biopsies were negative. Histologic study of the splenic lesion demonstrated low-grade malignant lymphoma: follicular mixed, small cleaved and large cell (the remaining splenic parenchyma showed a normal histologic pattern). Immunohistochemical analysis showed B-cell lymphoma and tumor cells expressed  $\kappa$ -immunoglobulin light chains.

The patient recovered from the splenectomy and she is currently well and in complete remission seven months after diagnosis; she did not receive chemotherapy postoperatively.

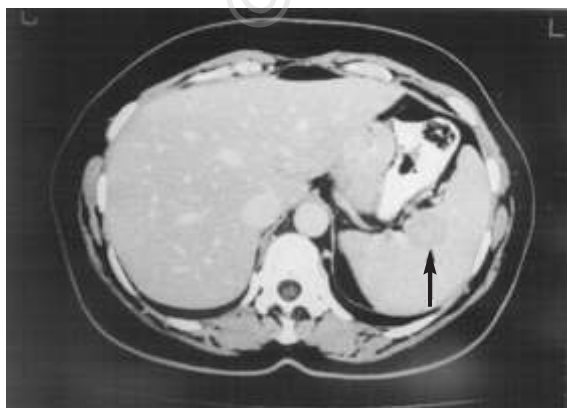


Figure 1. CT scan of the spleen demonstrating a normal-sized spleen with a focal low-density lesion, 2.5 cm in diameter (arrow).

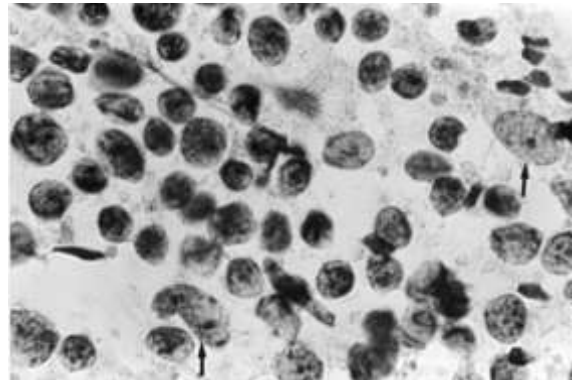


Figure 2. Ultrasonically-guided fine-needle aspiration biopsy of the focal splenic lesion shows a monomorphic pattern of atypical lymphoid cells consistent with NHL. The large cells (arrows) are endothelial cells of the splenic pulp (MGG staining x 400).

### Discussion

It is difficult when reviewing the literature to determine the actual frequency of primary splenic lymphoma because a standard set of criteria has not been used uniformly for this diagnosis. There are considerable variations in the extent of extrasplenic disease (i.e. lymph nodes, liver and bone marrow) at presentation in the different reports,<sup>7-11</sup> and thus the frequency of this entity varies depending on the criteria used by different investigators.<sup>2-4,7-11</sup>

Primary lymphoma of the spleen is very rare if the strict criteria for diagnosis suggested by Das Gupta *et al.*<sup>4</sup> are applied. Ahmann *et al.*<sup>7</sup> reported on a series of 49 cases in which the diagnosis of NHL was made at splenectomy; however, 32 of 49 patients had lymphomatous involvement of the liver or abdominal lymph nodes. Skarin *et al.*<sup>8</sup> reported 11 patients with splenic lymphoma, 9 of whom had periportal and/or sinusoidal malignant infiltration of the liver. Kraemer *et al.*<sup>9</sup> in their series of 49 cases with PSL included patients with liver and lymph-node involvement. Similarly, other studies<sup>10,11</sup> included cases with liver, bone marrow or abdominal lymph-node involvement as PSL.

On the other hand, Brox *et al.*<sup>2</sup> recently reported 9 patients with NHL that fulfilled the strict criteria for diagnosis of PSL suggested by Das Gupta *et al.*<sup>4</sup>

It is interesting that all the patients reported with PSL<sup>2-4,7-11</sup> showed splenomegaly, and a diag-

nosis of PSL was made by splenectomy. However, as reported by Gobbi *et al.*,<sup>3</sup> a resection of the spleen is not necessarily performed unless particular clinical problems are encountered. For this reason PSL is often diagnosed late, when it has spread to other organs, and therefore it is obviously difficult to determine the actual frequency of PSL that fulfill Das Gupta's criteria.

Our case was limited to the spleen and met the criteria proposed by Das Gupta *et al.*<sup>4</sup> It was a low-grade NHL type, like the majority of PSL cases reported.<sup>3</sup> It is interesting that in this case the spleen was not palpable, in contrast with previously reported cases,<sup>2-4,7-11</sup> and a focal lesion of the spleen was disclosed by US and CT.

We believe this to be the first report of a patient with PSL diagnosed by fine-needle guided biopsy. This case demonstrates the feasibility of so diagnosing primary splenic lymphoma, and that splenectomy may be not necessary for this diagnosis.

Non-invasive imaging techniques such as US and CT are useful in documenting splenomegaly and can adequately visualize focal nodules in the spleen less than 1 cm in diameter.<sup>6</sup>

The detection of focal defects in the spleen at US or CT examination may suggest the presence of lymphoma, and a definite diagnosis can be made by guided biopsy as previously reported.<sup>5,6</sup>

US-guided fine-needle aspiration biopsy cytologic examination has been shown to be useful in the diagnosis of splenic involvement by lymphoma.<sup>5</sup> In the spleen, however, malignant lymphocytes obtained in this manner can be distinguished from normal lymphocytes only with difficulty, and histologic confirmation may be required.<sup>6</sup> This is possible using US-guided tissue core biopsies that allow histologic and immunologic classification.<sup>6</sup>

The case described here demonstrates the feasibility of diagnosing primary splenic lymphoma in this manner. Early definite diagnosis is significant since early stage carries a favorable prognosis if appropriately treated.

Splenectomy is considered the most effective therapy for patients with PSL; however, whether splenectomy alone can be considered curative is as yet unresolved,<sup>2</sup> and adjuvant therapy in some subgroups of patients with PSL may also play an important role.<sup>2</sup>

Nevertheless, splenectomy has been performed in these patients with a double objective: a diagnostic one and a therapeutic one.<sup>2-4,7-11</sup> Since the diagnosis of PSL can be made by US-guided biopsy, as reported in our case, therapeutic procedures other than splenectomy, such as chemotherapy and/or radiotherapy, could be considered as first line treatment. Thus PSL patients could be better defined and the roles of surgery, chemotherapy and radiotherapy could be evaluated.

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