Case Report

Surgery Section

Primary Splenic Lymphoma: A Rare Clinical Case Report

HANUMANTHAPPA M.B., GOPINATHAN S., PRAVEEN NAYAK ,GURUPRASAD RAI D., SANKETH SHETTY

ABSTRACT

Primary Splenic Lymphoma (PSL) is a rare neoplasm of the spleen, probably comprising less than 2% of all the lymphomas [1, 7] and 1% of all the non-Hodgkin's lymphomas (NHL) [3]. The true incidence of the disease is difficult to estimate because of the variable definitions of the disease. Das Gupta et al. proposed that the diagnosis of PSL could be made when the disease was confined to the spleen or the hilar lymph nodes and that no recurrence of the disease was evident for at least 6 months after splenectomy [7]. The case which is reported here met these strict criteria. The patient is in complete remission one year after the diagnosis. Splenectomy is considered as the most effective therapy for PSL [2, 3, 6, 7]. In this article, we present a rare case of PSL in a 65 years old male. The histopathological evaluation of the splenectomy specimen revealed low grade NHL. The purpose of writing this article was to report this rare occurrence of PSL.

Key Words: Splenomegaly, Primary splenic lymphoma, Non-Hodgkin's lymphoma, Splenectomy

INTRODUCTION

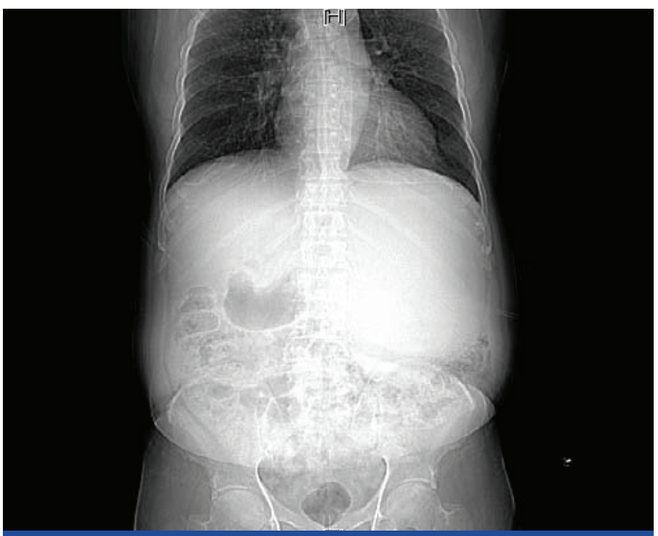
PSL is a rare condition which occurs in less than 1% of all the non-Hodgkin's lymphomas [7, 8].The spleen may be the primary site of the lymphoma or it may be an element of disseminated malignancy [7]. PSL is difficult to diagnose, as there is no standard definition and diagnostic criteria for it. Das Gupta et al. proposed that this diagnosis could be made when the disease was confined to the spleen or the hilar lymph nodes. No recurrence of the disease is evident for at least 6 months after splenectomy [3]. It usually presents with non specific symptoms like left upper quadrant pain or discomfort due to splenomegaly, pyrexia, weight loss, night sweats, and weakness. In this article, we are reporting a case of PSL in a 65 years old man who presented with splenomegaly and non-specific symptoms.

CASE REPORT

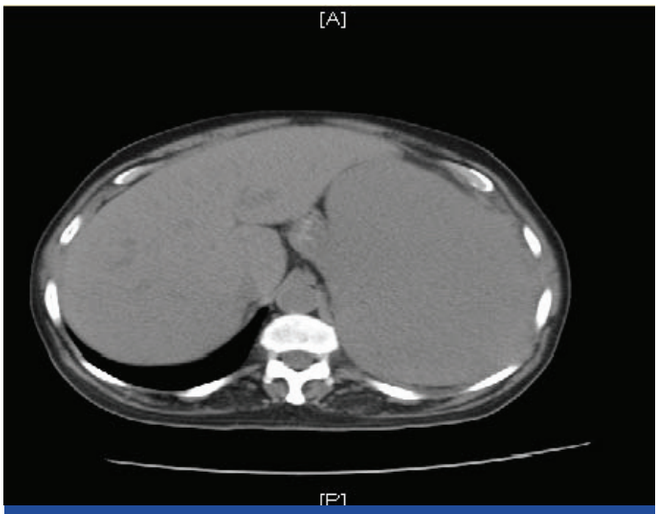
A 65 years old man presented with a two months history of abdominal pain, a mass in the abdomen, weakness and weight loss. He had no history of fever or night sweats. His abdominal examination revealed a large splenic mass. There was no hepatomegaly and abdominal or peripheral lymphadenopathy. The haemogram showed Hb of 8g/dl, a total count of 10,900 (N- 40%, L-55%, E- 4%, M-1%) and a platelet count of 1, 20,000 cells/cumm. His peripheral blood smear showed normocytic normochromic anaemia without any abnormal or atypical lymphocytes. Hid blood was negative for the malaria parasite. USG and CT abdomen revealed a large splenomegaly with cholecystolithiasis. There was no splenic hilar or abdominal adenopathy. His CT chest was within the normal limits. Lymphoma and tropical splenomegaly were considered in the differential diagnosis. An exploratory laparotomy was performed, which showed gross splenomegaly without any evidence of liver or abdominal lymph node involvement. An accessory spleen was not present. Splenectomy and cholecystectomy were done. The spleen measured 38x25x15cm and it weighed 3560 grams. The capsule was smooth and intact. The histopathological examination of the spleen revealed B cell type, low grade non-Hodgkin's lymphoma. There was no splenic hilar adenopathy. The bone marrow aspiration study did not reveal any evidence of lymphoma. Immunohistochemical studies revealed features which were suggestive of splenic lymphoma of the B cell type. He did not receive adjuvant chemotherapy post-operatively. The patient is in complete remission one year after the diagnosis.

DISCUSSION

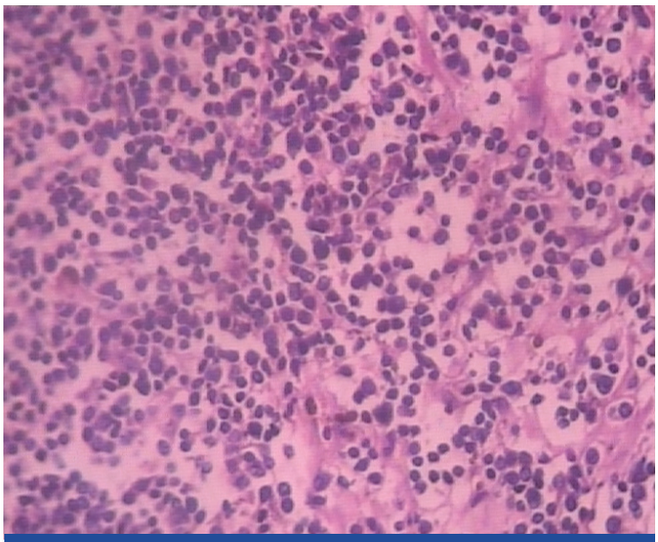
PSL is a rare condition which occurs in less than 1% of the non-Hodgkin's lymphomas [7, 8]. The spleen may be the primary site of the lymphoma or it may be a part of a disseminated malignancy [7]. The actual frequency of PSL is difficult to estimate because of the lack of a standard set of diagnostic criteria [3]. Some authors consider PSL to be an entity which is limited to the involvement of spleen and the splenic hilum [2]. Others consider it to be an entity that develops in the spleen and that has got the potential



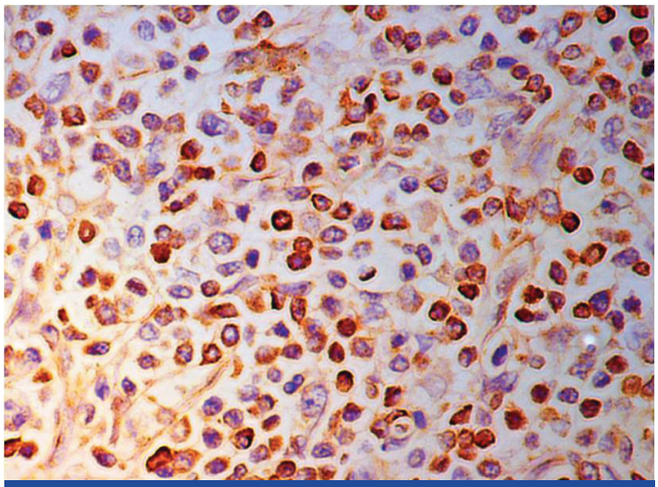
[Table/Fig-1]: C T scan of chest and upper abdomen



[Table/Fig-2]: C T scan abdomen showing gross splenomegaly



[Table/Fig-3]: Photo microgram showing features of lymphoma(under high power with Giemsa stain)



[Table/Fig-4]: Immunohistochemistry features suggesting Splenic Lymphoma ,B cell type

for spreading to other organs [2]. Das Gupta et al. proposed that the diagnosis of PSL can be made when the disease is confined to the spleen or the hilar lymph nodes, and that no recurrence of the disease was evident for at least 6 months after splenectomy [3]. Our case was confined to the spleen and it fulfilled the criteria which was proposed by Das Gupta et al. Usually, it presents with non-specific features like left upper quadrant pain, anorexia, weight loss, fever, night sweats and a mass in the abdomen. Cytopaenia can also be a presenting feature [7]. Rarely, it may be asymptomatic. The complete blood count and the peripheral blood smears are usually normal [6]. ESR may be elevated [7]. The diagnostic imaging techniques (USG/CT) are indispensible tools [6]. The most common appearance of PSL is hypodense splenic lesions on contrast enhanced CT scans or hypoechoic lesions on USG [7]. CT scan is also very helpful in excluding the involvement of abdominal adenopathy and other organ involvement. MRI is also a useful tool for identifying and characterizing the focal splenic lesions [7]. USG/CT guided percutaneous needle biopsy should be carefully considered, because it is associated with major complications in 1% of the cases and because its diagnostic accuracy is about 90% [6]. Immunohistochemistry is useful in detecting the cell type. On immuno-histochemical staining with the CD 20 antibody, it has been found that the splenic B-cell lymphomas are usually positive for CD 20. These lymphomas are usually of the NHL type, originating from the B cells [6]. The staging of PSL has been described by Ahmann and Kiely [7]. Stage 1 refers to the disease which is confined to the spleen; stage 2 implies the involvement of the spleen and the splenic hilar lymph nodes and stage 3 refers to the extra-splenic nodal or hepatic involvement [7]. The survival of the PSL patients significantly correlates to the stage of the disease [8]. Das Gupta et al. have described two types of PSL depending on the histology: low grade, lymphocytic lymphoma (well differentiated small cell variant or with lymphoplasmocytic/lymphoplasmocytoid differentiation), and intermediate grade lymphoma (diffuse or nodular mantle zone lymphoma) [1]. Our case was of the low grade NHL type, like the majority of PSL cases which were reported. Brox A et al., in their series of 9 cases of PSL, did not find any correlation between the histological subtype and the prognosis [5]. The possible treatment modalities include splenectomy, local radiotherapy and systemic chemotherapy [7, 8]. Splenectomy is considered as the most effective therapy for patients with PSL, because it is both a diagnostic and a therapeutic one [1, 2, 3, 9]. Local radiotherapy is considered for those in whom splenectomy is not feasible [7, 8]. Musteata et al. suggested that early splenectomy along with combination chemotherapy was the optimum treatment due to the higher rates of remission, a more prolonged duration of the remission and the better overall survival rates as compared to splenectomy or splenectomy with single agent CT [7]. The CHOP regimen appears to be the most widely accepted chemotherapy regimen for PSL [7].

CONCLUSION

PSL is a very rare condition, often presenting with non-specific features and a splenic mass. Most often, the diagnosis is made after the histopathological evaluation of the splenectomy specimen. A thorough clinical examination and investigation are mandatory to rule out the disease in other organs, including the liver, bone marrow and the lymph nodes. Splenectomy is the most effective therapy for Primary Splenic Lymphoma.

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AUTHOR(S):
1. Dr. Hanumanthappa M.B.
2. Dr. Gopinathan S.
3. Dr. Praveen Nayak
4. Dr. Guruprasad Rai D.
5. Dr. Sanketh Shetty

PARTICULARS OF CONTRIBUTORS
1. MBBS, MS Surgery
   Associate Professor, Department of Surgery,
   A J Institute of Medical Sciences, Mangalore
2. MBBS, MS Surgery
   Assistant Professor, Department of Surgery,
   A.J. Institute of Medical Sciences, Mangalore
3. MBBS, MS, Professor, Department of Surgery,
   A. J. Institute of Medical Sciences, Mangalore
4. MBBS, Resident, Department of Surgery,
   A. J. Institute of Medical Sciences, Mangalore
5. MBBS Resident, Department of Surgery,
   A. J. Institute of Medical Sciences, Mangalore

PLACE OF STUDY:
AJ Institute of Medical Sciences, Mangalore.

NAME, ADDRESS, TELEPHONE, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. M.B.Hanumanthappa
Dept of Surgery
AJ Institute of Medical sciences, Kuntikana
Mangalore, South Canara 575004, India.
Phone: 9845170266
Email: mb_h@rediffmail.com

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