Primary splenic lymphoma presenting with ascites

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Abstract

An 84 year-old gentleman presented with abdominal distension, anorexia and occasional epigastric pain over a four-week period. Blood parameters revealed a hypochromic microcytic anaemia. Both CT and US scan identified ascites and a mass in the left upper quadrant. An ascitic tap was performed identifying bloody ascites and the presence of reactive mesothelial cells on cytology. A subsequent laparotomy and splenectomy was performed. Histology of the resected spleen revealed a Grade 2 follicular lymphoma (Figure 2). The patient had an uneventful postoperative recovery and was well at 6 months follow up. The spleen is an organ with an important immunological function. Primary splenic involvement occurs in less than 1% of non-hodgkin's lymphoma. Symptoms of primary splenic lymphoma (PSL) include pyrexia, weight-loss, night sweats, generalised weakness and left upper quadrant pain secondary to splenomegaly. Ascites is a rare presenting feature of PSL. This report illustrates a case of primary splenic lymphoma which poses diagnostic challenges for the pathologist and clinician and ultimately requires definitive splenectomy to confirm a diagnosis.

Introduction

Primary splenic lymphoma (PSL) is a rare condition occurring in less than 1% of non-Hodgkin's lymphoma. PSL often poses a diagnostic dilemma and symptoms include pyrexia, weight-loss, night sweats, generalised weakness and left upper quadrant pain secondary to splenomegaly. This case report highlights a case of primary splenic lymphoma in an elderly gentleman and illustrates the management of such a case.

Case Report

An 84 year-old gentleman presented to the emergency department with a four-week history of abdominal distension and associated anorexia with occasional epigastric pain and dysnpoea. Of note, he had a background history of rheumatoid arthritis. On examination his abdomen was soft but distended with ascites. No abdominal masses were palpable and the examination was otherwise normal.

Bloodwork revealed a hypochromic, microcytic anaemia, with a slightly elevated CRP and LDH. Bone marrow biopsy and myeloma screen were negative. Trans-thoracic echo was essentially normal with an ejection fraction of approximately 50%. A colonoscopy was performed and showed sigmoid diverticular disease. Abdominal ultrasound demonstrated ascites and a 10 cm mass in the left upper quadrant related to the upper border of the spleen. A subsequent CT scan also identified ascites and a left upper quadrant mass with celiac lymphadenopathy and bilateral pleural effusions (Figure 1).

An ascitic tap was performed and yielded a bloody aspirate. The albumin level of the ascites was 29 g/L, with a total protein content of 52 g/L, fluid LDH of 303 U/L and amylase of 15U/L. Cytology identified reactive mesothelial cells and a cell-block yielded a predominantly epithelial cell population. At this stage, the patient began to experience worsening abdominal discomfort due to the increasing abdominal distension. A laparoscopy was performed, at which an enlarged spleen and copious bloody ascites was noted. No active haemorrhage was identified and there was no evidence of peritoneal or omental disease. All other intra-abdominal viscera were normal. Three litres of ascitic fluid was drained in an effort to obtain definitive cytology. Again, only reactive mesothelial cells were identified. The ascitic fluid reaccumulated within one week, once again causing severe abdominal discomfort and following a full discussion with the patient and his family, an elective splenectomy was planned. At laparotomy, a large splenic mass, involving the left crus, left diaphragmatic dome, stomach and splenic flexure of the colon was visualised. The mass was completely resected and three litres of bloody ascitic fluid was drained.

The resected spleen weighed 585 g and measured 140 mm × 100 mm × 100 mm. Histology showed the splenic parenchyma to be effaced by a lymphoid infiltrate, which was CD20 positive, indicative of a B cell lineage. CD10 was also positive. The appearances were consistent with a low-grade, non-Hodgkin's B-cell follicular lymphoma, with a focus of transformation to high-grade lymphoma or possibly a composite lymphoma (i.e. non-Hodgkin's plus Hodgkin's lymphoma). Following expert second opinion, a final diagnosis of Grade 2 follicular lymphoma with areas of Grade 3b cytology was confirmed. The patient made an uneventful recovery and was discharged eight days post-operatively, without reaccumulation of the ascites. He was entirely well at six-month out-patient review and refused any form of adjuvant treatment.

Discussion

Primary Splenic lymphoma (PSL) is a rare condition, representing less than 1% of non-Hodgkin's lymphoma (NHL).[1] The spleen may be the primary site of the lymphoma or it may be an element of disseminated malignancy. PSL can be difficult to diagnose as half of all patients with Hodgkin's Disease and one third of those with non-Hodgkin's lymphoma have splenic involvement. Similarly, the definition of PSL is somewhat controversial. The case we report corresponds with the definition by Das Gupta et al, who describe PSL as a lymphoma only involving the spleen and hilar lymph nodes, but not the liver or other sites.[2] In order for the diagnosis to be confirmed there must be a six-month relapse-free period following splenectomy. Definitions vary, however. Skarin et al suggest that PSL is any lymphoma with splenic involvement in which splenomegaly is the dominant feature.[3] Symptoms of PSL include pyrexia, weight-loss, night sweats, generalised weakness and left upper quadrant pain secondary to splenomegaly. Cytopenia can also be a presenting feature and significant laboratory findings include elevated ESR and β2 microglobulin.[4] The principal clinical finding is splenomegaly, which in this partic-

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ular case was impossible to demonstrate, due to the gross ascites. To our knowledge, this is the first reported case of PSL presenting with ascites. The most common appearance of PSL on diagnostic imaging studies is of hypodense splenic lesions on contrast enhanced CT scans or hypoechoic lesions on ultrasound.[5] Contrast enhanced ultrasound may also prove useful in evaluating splenic lesions.[6] MRI is also a useful tool for identifying and characterizing focal splenic lesions.[7] The differential diagnosis for such appearances is broad and splenectomy is often required for definitive diagnosis. Kraus *et al.* demonstrated that whilst malignancy is the most common diagnosis in patients with a splenic mass, splenectomy is frequently refused by patients and in such cases radiologically-guided, percutaneous biopsy may offer an alternative modality to diagnose and histologically subtype splenic lymphoma.[8] Disease staging of PSL has been described by Ahmann and Kiely.[9] Stage I refers to disease confined to the spleen; Stage II implies involvement of the spleen and hilar lymph nodes and Stage III refers to extra-splenic nodal or hepatic involvement. 5-year survival is relatively poor, with a 31% overall survival for all groups.[9] Kehoe *et al.,* with a series of 21 patients over a 5-year period, demonstrated a combined survival rate of 43% for Stage I and II disease, but just 14% for Stage III.[10] However, as both of these studies are quite old, there is a need for an updated study to take into account novel imaging modalities and therapeutic options. Xiros *et al.* maintain that the median survival after diagnostic splenectomy is 24 months regardless of disease stage or adjuvant chemotherapeutic regimen.[11] Treatment for PSL sparks debate due to the lack of clinical trials. Possible treatment modalities include splenectomy, local radiotherapy and systemic chemotherapy. Splenectomy is often both diagnostic and therapeutic. Local radiotherapy is considered for those in whom splenectomy is not an option. Morel *et al.* stated that early splenectomy can improve survival for those patients in whom cytopenia improves after surgery and those who tolerate adjuvant chemotherapy.[4] Musteata *et al.* after looking at 104 patients with primary low grade NHL of the spleen concluded that early splenectomy along with combination chemotherapy is the optimum treatment due to higher rates of remission, a more prolonged duration of remission and better overall survival rates when compared with splenectomy or splenectomy with single agent chemotherapy.[12] The CHOP regimen appears to be the most widely accepted chemotherapy regimen for PSL, according to the individual cases despite the lack of clinical trials on the subject.

Conclusion

In summary, PSL is a rare condition which can be difficult to diagnose. Splenectomy offers both definitive histological diagnosis and therapeutic resection and represents the mainstay of treatment. Adjuvant chemotherapy and radiotherapy may be of use and outcomes, unsurprisingly, are far superior with early stage (I & II) disease. When splenectomy is combined with combination chemotherapy, outcomes in terms of remission and survival are improved.

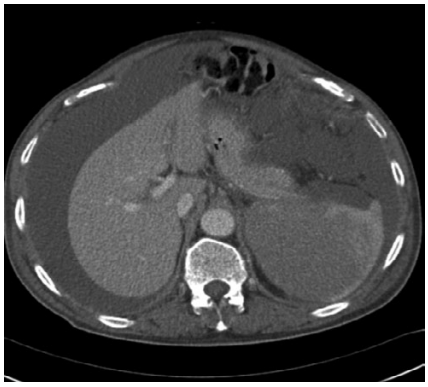

Figure 1. Computed tomography of the abdomen showing an enlarged spleen and the presence of ascites

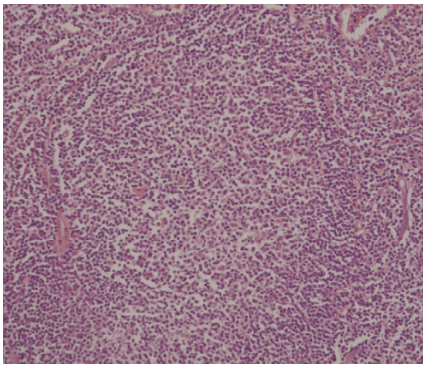

Figure 2. Photograph of histology slide displaying the lymphoma at 10X magnification.

References

1. Brox A, Shustik C. Non-Hodgkin's lymphoma of the spleen. Leuk Lymphoma, 1993;11:165-71.
2. Dasgupta T, Coombes B, Brasfield RD. Primary Malignant Neoplasms of the Spleen. Surg Gynecol Obstet 1965;120:947-60.
3. Skarin AT, Davey FR , Moloney WC. Lymphosarcoma of the spleen. Results of diagnostic splenectomy in 11 patients. Arch Intern Med, 1971;127:259-65.
4. Morel P, Dupriez B, Gosselin B, et al. Role of early splenectomy in malignant lymphomas with prominent splenic involvement (primary lymphomas of the spleen). A study of 59 cases. Cancer 1993;71:207-15.
5. Dachman AH, Buck JL, Krishnan J, et al. Primary non-Hodgkin's splenic lymphoma. Clin Radiol 1998;53:137-42.
6. Sutherland T, Temple F, Hennessy O, Lee WK. Contrast-enhanced ultrasound features of primary splenic lymphoma. J Clin Ultrasound 2010;38:317-9.
7. Ioannidis O, Papaemmanouil S, Paraskevas G, et al. Pathologic Rupture of the Spleen as the Presenting Symptom of Primary Splenic Non-Hodgkin Lymphoma. J Gastrointest Cancer 2011 Apr 26. [Epub ahead of print]
8. Kraus MD, Fleming MD, Vonderheide RH. The spleen as a diagnostic specimen: a review of 10 years' experience at two tertiary care institutions. Cancer 2001;91: 2001-9.
9. Ahmann DL, Kiely JM, Harrison EG Jr, Payne WS. Malignant lymphoma of the spleen. A review of 49 cases in which the diagnosis was made at splenectomy. Cancer 1966;19:461-9.
10. Kehoe J, Straus DJ. Primary lymphoma of the spleen. Clinical features and outcome after splenectomy. Cancer 1988;62:1433-8.
11. Xiros N, Economopoulos T, Christodoulidis C, et al. Splenectomy in patients with malignant non-Hodgkin's lymphoma. Eur J Haematol 2000;64:145-50.
12. Musteata VG, Corcimaru IT, Iacovleva IA, et al. Treatment options for primary splenic low-grade non-Hodgkin's lymphomas. Clin Lab Haematol 2004;26:397-401