Multilobated B-cell lymphoma of the spleen

D.J. Farrell, C.A. Bloxham and H.H. Lucraft

Department of Histopathology and 'Department of Clinical Oncology, Newcastle General Hospital, Newcastle upon Tyne, UK

Summary: We report a case of multilobated B-cell lymphoma presenting with primary splenic involvement. This is a very unusual tumour occurring in an uncommon site.

Introduction

Peripheral T-cell lymphomas characterized by cells having large multilobated nuclei were first described by Pinkus et al. in 1979. Subsequently multilobated non-Hodgkin's lymphomas with a B-cell phenotype were reported. These tumours may occur in nodal, extranodal or combined locations but most frequently present in an extranodal site. We report a case presenting with hypogammaglobulinaemia and splenomegaly. This is a very rare occurrence.
Case history

A 37 year old man was found to have hypogammaglobulinaemia in 1989 following investigation for recurrent chest infections. He began treatment with weekly immunoglobulin injections. In July 1990 he developed severe pain in his left posterior lower chest and drenching night sweats. He was found to have splenomegaly. Computed tomographic (CT) scan confirmed that the spleen was enlarged and contained multiple soft tissue non-enhancing nodules and also showed multiple enlarged lymph nodes in the mesentery and around the origin of the superior mesenteric artery. The liver appeared radiologically normal. CT scan of the chest showed no evidence of disease and there was no palpable lymphadenopathy. Diagnostic laparotomy and splenectomy was performed in November 1990. Preoperative blood count was normal. Bone marrow aspirate and trephine showed reactive hypocellular marrow with no evidence of lymphoma. Lactate dehydrogenase was raised at 759 U/l (normal less than 430 U/l).

A clinical diagnosis of Stage IIB high-grade non-Hodgkin's lymphoma was made and he was treated with combination chemotherapy (cyclophosphamide, vincristine, adriamycin and prednisolone in pulsed courses). He was in complete remission at completion of chemotherapy in March 1991. He has subsequently remained well with no evidence of further disease (to February 1993). Hypogammaglobulinaemia has persisted and he continues to receive gammaglobulin replacement.

Pathological findings

Macroscopic examination showed a spleen weighing 530 g and measuring 16 x 11 x 5 cm. The surface of the spleen was nodular. On sectioning there were many white tumour nodules which were irregular in outline but well demarcated and measuring up to 2.5 cm in maximum dimension. Within the splenic hilum and the attached omentum there were several small lymph nodes measuring up to 1 cm in diameter which had a grey cut surface. Microscopic examination showed that the tumour deposits were composed of large pleomorphic lymphoid cells with vesicular nuclei, prominent large round eosinophilic nucleoli and variable amounts of eosinophilic cytoplasm (Figure 1), which comprised up to 53% of the malignant infiltrate. The nuclei of many tumour cells were markedly irregular with deep clefts and an occasional clover leaf pattern (Figure 2) characteristic of the multilobated lymphoma. Scattered multinucleated cells were also present but classical Reed-Sternberg cells were not identified. Foci of necrosis and fibrosis were present throughout and in areas only a perivascular ring of viable tumour cells was seen. A T-cell lymphoma was thought most likely on morphological grounds but immunohistochemistry of the tumour cells showed positive staining using the monoclonal antibodies MB1 (CD45RA), MB2 and L26 (CD20), and negative staining for MT1 (CD43) and UCHL1 (CD45RO). Staining for CD30 and CD15 was also negative. Thus the immunohistochemical profile was consistent with B-cell phenotype lymphoma. The lymph nodes recovered from the splenic hilum and omentum were not involved by the tumour which suggests that the malignancy may have been confined to the spleen. However, the enlarged mesenteric lymph nodes noted on CT scan were not biopsied.

Discussion

Multilobated non-Hodgkin's lymphomas are a well recognized though uncommon entity. The
original case reports suggested that these were T-cell lymphomas but subsequent phenotyping has shown that both B-cell and T-cell varieties exist.

Several series of multilobated B-cell lymphoma have been reported and these tumours, similar to their T-cell counterparts, present preferentially as extranodal lesions. However, splenomegaly is a very rare presenting feature. Van Baarlen et al. presented 30 and nine cases, respectively, in which this tumour involved a wide variety of tissues such as parotid, maxilla, soft tissues, bone and testis among others, but none of these cases showed splenic involvement. Indeed, to our knowledge there has only been one previous case report of multilobated B-cell lymphoma presenting with primary splenic involvement, making this case a very unusual site of presentation for this unusual tumour. It is important to recognize this rare morphological subtype, particularly when arising in an extranodal site, as prognosis for multilobated lymphomas appears to be rather favourable with a striking incidence of long-term remission even after inadequate local therapy.

Hypogammaglobulinaemia is seen to a greater or lesser extent in all B-cell neoplasms. Therefore, if hypogammaglobulinaemia is discovered in any patient above the age of 20 which cannot be explained through other causes, it should be taken to indicate B-cell neoplasia until proved otherwise.

It is suggested that in B-cell neoplasms physiological regulators of antibody responses (possibly suppressor T-cells) are activated, which selectively moderate B-cell triggering by antigen. Other suggested possible mechanisms are increased catabolism of immunoglobulins and induced hypersplenism with consequent antigen capture and breakdown. The reduced immunoglobulin levels impair primary antibody responses more profoundly than secondary responses and patients are susceptible to pyogenic infections, especially chest infections, as occurred in this case.

In summary, multilobated lymphomas are a distinct morphological subtype which may be of B-cell or T-cell origin. They often present in extranodal locations and have a good prognosis. The phenotype of these large cell lymphomas cannot be determined with certainty by the morphological appearances of the tumour cells and immunocytochemistry is required for a definite diagnosis of phenotype. This case is unusual in that this is an uncommon type of lymphoma presenting in an unusual manner and which appears to be confined to the spleen.

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References


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