Primary splenic lymphoma with filiform ultrastructure

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Abstract

A case of primary large cell splenic lymphoma of B lineage exhibiting filiform cell appearance is reported. The patient presented with massive splenomegaly, and following spontaneous splenic rupture, died of adult respiratory distress syndrome. The clinical aspects of the case, notably a lymphoma arising as a primary tumour in the spleen, with spontaneous spleen rupture and rapid fatal outcome, in combination with the filiform appearance of the lymphoma on electron microscopic examination, constitute an unusual combination of features. As far as is known, this B cell neoplasm is only the second primary splenic lymphoma of filiform type to be recorded.

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Although secondary disease in the spleen as a result of lymphoma is common, primary splenic lymphomas presenting without blood, marrow, or nodal disease occur in less than 1% of patients with lymphoma.¹ The histological types recognised include: diffuse small cell lymphocytic; diffuse large cell; diffuse small cleaved cell; diffuse large cell immunoblastic; follicular small cleaved cell; follicular mixed small and large cell²; and large cell anaplastic Ki-1 type.³ The entity known as splenic lymphoma with villous lymphocytes has also been described,^{4,5} but here there is peripheral blood disease. Recently the filiform cell appearance in a large cell primary splenic lymphoma was documented for the first time.⁶ The aim of this report is to document what we believe is only the second case of such a tumour. It differs in some important clinical aspects from the first recorded example.

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Case report

A 65 year old Chinese man, who had lived in the United Kingdom for 30 years, was admitted to hospital with chest infection associated with dyspnoea and pleuritic pain. On examination he was noted to have hepatosplenomegaly and was pancytopenic. A bone marrow aspirate showed reactive features. There were no villous lymphocytes or hairy cells in the peripheral blood. Lymph nodes were not enlarged. Two days after admission he developed an acute intra-abdominal bleed. At laparotomy the spleen was seen to have ruptured and a splenectomy was carried out. The patient remained unwell, developed adult respiratory distress syndrome, and died two weeks later. A post mortem examination was not carried out.

Pathology

Tissue from the splenectomy specimen was fixed in 10% neutral buffered formalin and embedded in paraffin wax. Sections 4-5 µm

Figure 1 (A) Section showing rounded and (arrowhead) pleomorphic lymphoid cells infiltrating sinuses. (haematoxylin and eosin). (B) Immunoreactivity for the B cell marker MB2 (arrowheads) (ABC method)

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Figure 2 (A) Tumour cells showing rounded and irregular nuclei. In the cytoplasm note lipid (L) and clusters of mitochondria (m). Arrowheads indicate areas of intercellular space filled with cell processes. (B) Detail of microvillous processes in longitudinal (arrowheads) and cross-section (arrows).

thick were stained with haematoxylin and eosin and for chloroacetate esterase. Additional sections were stained using the avidin-biotin-peroxidase complex method with the following antibodies: anti-leucocyte common antigen (Dako UK; 1 in 20 dilution); anti-epithelial membrane antigen (Dako; 1 in 50); MB2 (Biotest UK; 1 in 20); L26 (Dako; 1 in 100); anti-CD30 (Ber-H2) (Dako; 1 in 20); anti-CD3 (Dako; 1 in 200); anti-CD43 (Dako; 1 in 50); Mac387 (Dako; 1 in 100) and KP1 (CD68) (Dako; 1 in 100). For electron microscopy, tissue was retrieved from formalin and, after 1% osmium tetroxide and 0.5% uranyl acetate steps, was dehydrated and embedded in epoxy resin (Agar Scientific UK). Ultrathin sections were stained in 5% aqueous uranyl acetate and Reynolds' lead citrate.

The spleen was massively enlarged, measuring $26.0 \times 18.0 \times 8.5$ cm, and weighed 2850 g. There was extensive disruption of the capsule and underlying splenic pulp. Numerous infarcts of various sizes were present within the parenchyma.

In haematoxylin and eosin stained sections, there was diffuse infiltration of both red and white pulp by large, pleomorphic cells having

abundant eosinophilic cytoplasm and vesicular nuclei with multiple prominent nucleoli (fig 1A). Many bizarre nuclear forms were present and there were several multinucleated cells. Extensive necrosis was seen, and there were numerous normal and abnormal mitoses.

Tumour cells were positive only with the following antibodies: anti-leucocyte common antigen, MB2 (fig 1B), and L26 (the latter focally). Cells were negative for chloroacetate esterase.

At electron microscopy, nuclei varied in their degree of irregularity. Some had a regular outline, while many had small or deep invaginations (fig 2A). Well circumscribed, moderate to large, nucleoli were present, and there was modest peripheral heterochromatin. The cytoplasm contained focally numerous, artefactually distended mitochondria, some lipid, a few cisternae of rough endoplasmic reticulum, and polyribosomes. The cell surfaces were thrown up into numerous cell processes which filled up the intercellular space (fig 2B). These processes lacked the glycocalyx and the microfilament core rootlets typically found in glandular type microvilli. Junctions and external lamina were absent.

Discussion

On the basis of the histological, immunohistochemical, and electron microscopic results, this primary splenic tumour was a large cell, Ki-1 negative, non-Hodgkin's lymphoma of B lineage showing filiform ultrastructure. This tumour can be differentiated from the other two well known B cell neoplasms frequently affecting the spleen—namely, splenic lymphoma with villous lymphocytes,^{4,5} and hairy cell leukemia—by the small cell nature of the latter two conditions and the presence in peripheral blood of villous and hairy cells, respectively. Early on in the work-up of this

tumour, consideration was given to the possibility of a histiocytic neoplasm.⁷ The positive staining for B cell markers and negative staining for cells of macrophage lineage, however, failed to support this possibility. The tumour in this case lacked sufficiently distinctive histological and immunohistochemical features to be recognised as a filiform lymphoma, the definition of which, of course, depends on electron microscopic examination. It was none the less consistent in its immunophenotype (positive staining for leucocyte common antigen and B cell markers; negative staining for BerH2, T cell markers, epithelial membrane antigen, and Mac387 and CD68) with other filiform lymphomas.^{8,9}

The surface features shown by electron microscopy were reminiscent of the anemone cell appearance. This has been described in carcinomas, Hodgkin's disease, in a neuroendocrine tumour and in an ependymoma, although most cases—over 40—have been lymphomas (see Carstens for a recent concise review and references).¹⁰ Although the cell processes in our case were numerous, the typical anemone cell appearance in which cells are rather widely separated and in which the intercellular spaces are packed with processes was not quite seen in our case, and we therefore prefer to use the term filiform lymphoma rather than anemone cell lymphoma. Recently, a non-Hodgkin's splenic lymphoma was documented⁶ which, like that of the present report, was of B cell immunophenotype and of filiform ultrastructure, but which presented with quite different clinical features. Both cases were middle-aged to elderly men. Our case, however, presented with a massively enlarged spleen associated with hyper-

splenism, spontaneous splenic rupture, and early disseminated intravascular coagulation and infection, to which the patient finally succumbed within a short period after surgery. In the previous case⁶ the patient was reported as being well and free of recurrence 21 months after presentation.

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