Spleen pathology

Introduction

Open surgery for splenectomy is performed with diminishing frequency and pathology of this organ remains a poorly understood topic. Guided needle core biopsy is used increasingly to sample solid lesions within the spleen and, for a variety of pathologies, laparoscopic splenectomy is now the surgical method of choice. These changes in surgical practice influence the nature of splenic tissue samples with which pathologists require familiarity and, as in so many areas of practice, dialogue between professionals is important to ensure that appropriate material is obtained if histology is to be undertaken.

Needle biopsy offers a minimally invasive means of investigating spleen pathology and can be used safely in skilled hands to sample diffuse lesions. However, it is generally reserved for targeting relatively large, solid intrasplenic lesions, to minimize risk of haemorrhage. For the pathologist, needle core specimens pose similar challenges to those generated from other sites; only a small amount of tissue is available, often with a requirement to achieve numerous sections for a range of immunostaining, and background tissue is absent or minimal against which to assess the distribution and other architectural features of the sampled area. Also, despite use of ultrasound or computed tomography (CT) for guidance, splenic needle biopsy occasionally misses its target even in the hands of a skilled operator and yields uninformative background tissue. It should always be recommended that multiple cores of tissue are obtained. Tiny or highly fragmented specimens are, at best, suboptimal and may be non-diagnostic, with requirement for the procedure to be repeated.

Laparoscopic removal can be achieved safely for quite substantially enlarged spleens (500–800 g) and even larger ones under selected circumstances. Surprisingly large and intact pieces of tissue can be removed by this means if the surgeon is aware that this is necessary for pathological interpretation. However, most of the tissue is morcellated for removal and the resulting disrupted fragments are generally unsuitable for histological assessment; most cellular elements are stripped away from their supporting stroma. The smaller fragments consist predominantly of fragmented and tangled capsular and stromal connective tissue. Laparoscopic splenectomy is widely practised for removal of spleens from patients with hypersplenic symptoms accompanying mechanical red cell disorders and other sequestration syndromes (e.g., hereditary spherocytosis, auto-immune haemolytic anaemia, haemoglobinopathies, thalassaemias and idiopathic thrombocytopenic purpura). There is a perception, incorrect in at least a significant proportion of such patients, that histopathology has no importance in these circumstances. However, demonstration of an unexpected underlying condition or complication is not uncommon, in addition to the value of confirmation that the tissue shows an expected pattern of change for the patient's known clinical disorder.

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Questions

Case 1

A 40-year-old man presented with abdominal pain and anaemia. He was found to have massive splenomegaly, with no lymphadenopathy or other abnormal findings on CT scan. Splenectomy was performed. The spleen weighed 3000 g and had an intact capsule. It appeared irregularly nodular and slicing revealed prominent haemorrhagic and necrotic areas; no recognizable normal parenchyma was present. Histology varied widely between individual nodules and this variation is illustrated in Figures 1–3. What is the diagnosis/differential diagnosis?

Case 2

A 35-year-old man presented with abdominal discomfort. Fifteen years previously he had been treated for nodular lymphocyte-predominant Hodgkin lymphoma, with complete remission. Ultrasound and CT scans showed a substantially enlarged spleen with a diffusely nodular parenchyma, accompanied by para-aortic lymphadenopathy but no peripheral lymph node enlargement. Splenic biopsy (unusually, by a wedge biopsy procedure) was performed and a retroabdominal lymph node was also sampled by needle biopsy. Figures 4–6 show histological features of the white pulp-based nodules and intervening red pulp.

(a) What is the differential diagnosis?
(b) What immunohistochemistry would you do?

Case 3

A 67-year-old woman was diagnosed, one year previously, with low-grade B-cell lymphoma, consistent with splenic marginal zone B-cell lymphoma, on the basis of peripheral blood lymphocytosis and bone marrow involvement. She had persistent lymphocytosis and splenomegaly, despite chemotherapy, and splenectomy was undertaken since this can produce sustained partial or complete remission in some patients. The spleen weighed 550 g and measured 165 × 125 × 100 mm. The capsule was normal and the parenchyma appeared diffusely somewhat pale but otherwise normal; no focal abnormalities were evident macroscopically. Figures 7–9 illustrate the major histological features present. What is the differential diagnosis?

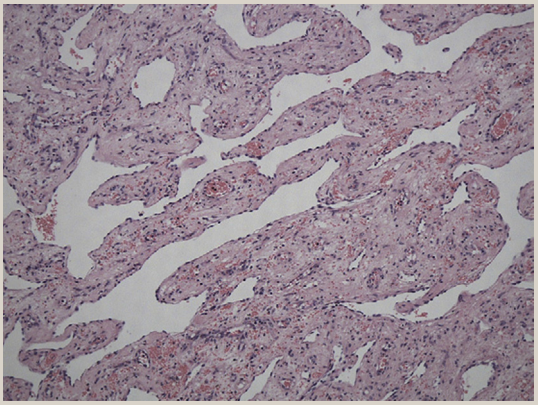

Figure 1

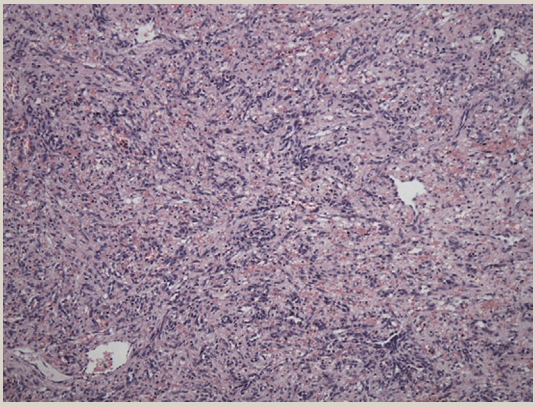

Figure 2

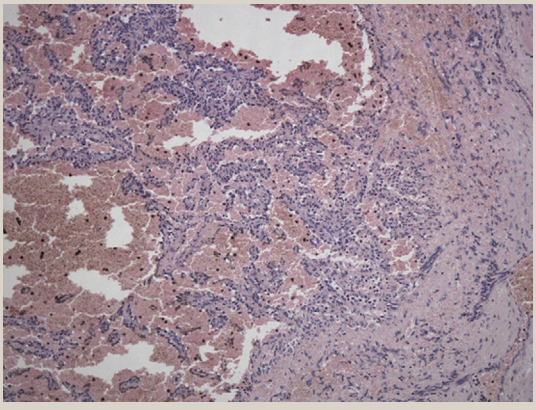

Figure 3

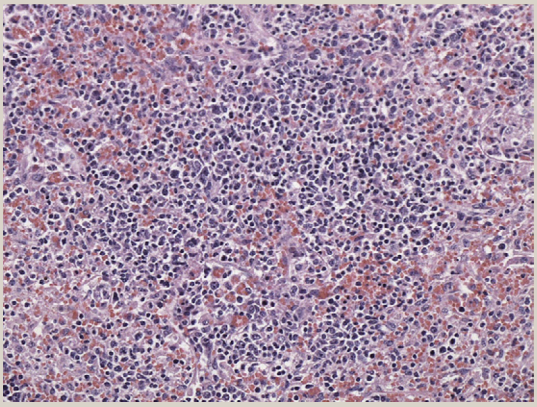

Figure 4

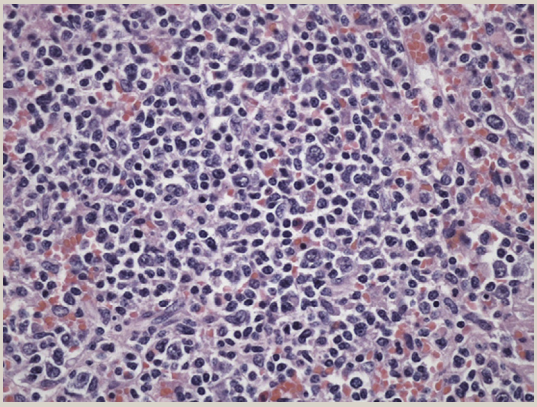

Figure 5

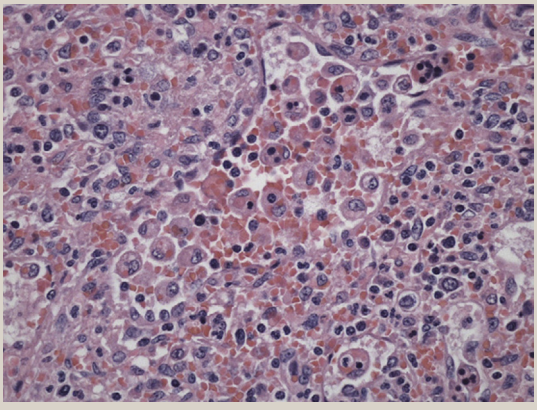

Figure 6

Answers

Case 1

Histology confirmed extensive replacement of the spleen by a multinodular mass with multiple areas of haemorrhage and necrosis. Different areas showed varying combinations of solid spindle cell and vasoformative growth. The latter included dense masses of closely packed capillary-like vessels, cavernous areas and cystic areas lined by papillary formations of littoral-type endothelial cells. Overt cellular atypia was minimal in all areas and no areas of high proliferative activity were identified by Ki67 immunostaining. Immunostaining for CD31 and CD34 was positive, with absent expression of the latter in areas of obvious littoral cell differentiation (Figure 10). Varying degrees of CD8 expression by endothelial cells (typical of littoral endothelium that lines normal splenic red pulp sinusoids) were found in all areas (Figure 11). No