Vascular neoplasms of the spleen

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Abstract

The spleen is an enigmatic organ that for a long time was thought to be useless, it is anything but. Made up of two main functional units, the spleen plays an important role in the immune system (white pulp) and in blood filtration and removal of unwanted material (red pulp).

Neoplasms of the spleen are either primary or secondary. Primary tumours can be lymphoid or non-lymphoid. This review is focused on the most common non-lymphoid tumours namely, the vascular neoplasm, which includes benign, malignant and intermediate grade tumours.

Keywords angiosarcoma; haemangioma; Kaposi sarcoma; littoral cell angioma; spleen; vascular neoplasm

Introduction

Vascular splenic neoplasms are the most common non-lymphoid primary tumour of the spleen.[1] The benign vascular tumours are: haemangioma, lymphangioma, and peliosis. Splenic lesions with intermediate biological behaviour include littoral cell angioma, Kaposi sarcoma and hemangioendothelioma. The main primary malignant splenic vascular lesion is angiosarcoma. See Table 1 for vascular tumours summary.

Haemangioma

These are the most common benign vascular splenic tumour. Autopsy series revealed a prevalence of up to 14%, mainly in adults.[2] These lesions are often asymptomatic and usually found incidentally. Most haemangiomas are small (less than 2 cm) and complication arises when they start to enlarge increasing the risk for spontaneous rupture.[2] Clinically they may present as a palpable mass with abdominal pain or discomfort, as hypersplenism with cytopenias or as consumption coagulopathy. Gross examination reveals well defined but non-encapsulated solitary or multiple blue-red spongy nodules. Solid larger lesions can be partly cystic. Calcifications, infarcts or fibrosis can be found in such larger lesions.[3] Histologically they are located in the red pulp and may be cavernous (most common) or capillary type[4] (Figure 1). They consist of interconnected vascular channels of variable size, ranging from capillary to cavernous, which are lined with a single layer of endothelium[5] (Figure 2). The endothelial cells are usually bland and flattened but, may be plump. Thromboses in these vascular channels are not uncommon. Cellular pleomorphism, nuclear hyperchromasia and mitosis (except in children) are absent.[6] Immunohistochemical stains reveal the endothelials cells positive for CD31, CD34 and factor VIII. CD8, CD68 and CD21 are negative. The latter three immunohistochemical stains are important in the differential diagnoses. Littoral cell angioma (LCA) is characterized by cuboidal or tall endothelial cells with papillary projections protruding into vascular spaces. LCA endothelial cells are positive for CD68 and CD21 and negative for CD34 and CD8. Peliosis is almost always associated with hepatic peliosis and is characterized by dilated sinuses with flattened lining cells that are positive for CD8 and CD68. Splenic hamartoma contain a mixture of unorganized vascular channels lined by endothelial cells and surrounded by fibrotic cords of predominant splenic red pulp with or without white pulp. Lymphangioma is most common in children and shows dilated cystic spaces containing proteinaceous material, lymph, and cholesterol clefts. Finally, hemangioendothelioma and angiosarcoma both show greater cytologic atypia.

Lymphangioma

Lymphangiomas are rare benign lesions that occur mostly in young patients.[4] The clinical picture ranges from asymptomatic incidental finding to symptomatic mass.[2] Splenic lymphangioma can be associated with lymphangiomatosis, a syndrome in which multiple organs including bone, soft tissue, or viscera are involved.[7] Grossly these present as solitary or multilocular cystic masses, than can be subcapsular if solitary. Histologically lymphangiomas can be divided into three types: capillary, cavernous, and cystic.[8] They are characterized by honeycomb of cystic spaces containing proteinaceous material, lymph-like or clear fluids and/or cholesterol clefts (Figure 3). These spaces are lined by a single layer of flattened endothelium (Figure 4). This layer may form small papillary projections. Immunohistochemical stains Factor VIII-related antigen and CD31 are the most reliable markers for the endothelial cells lining the cystic spaces. The main differential diagnosis is haemangioma. Both splenic lymphangioma and haemangioma are vasoformative tumours and they are closely related, but splenic lymphangioma is far less common and unlike the random localization seen with haemangiomas, lymphangioma often involves the capsule and trabeculae of the spleen, where lymphatics are normally concentrated.[1] Rare cases of splenic lymphangioma developing into malignant lymphangiosarcoma have been documented.[2]

Peliosis

Splenic peliosis are rare diseases, characterized by multiple blood-filled cystic spaces. These are usually associated with peliosis hepatis but cases of isolated splenic peliosis have been reported in the literature.[2] Most cases have an association with use of anabolic steroids; haematologic disorders such as aplastic anaemia; and wasting diseases such as tuberculosis, AIDS, and cancer.[2] They may also be associated with spontaneous splenic

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Summary of characteristic features of selected vascular tumour of spleen

Neoplasm	Clinical features	Histomorphologic data	Immunohistochemical findings
Haemangioma	Palpable mass with abdominal pain or discomfort	Interconnected vascular channels of variable size lined by bland endothelial cells	• Endothelials cells positive for CD31, CD34 and factor VIII • CD8, CD68 and CD21 are negative
Lymphangioma	Asymptomatic incidental finding to symptomatic mass	Cystic spaces containing proteinaceous material, lymph-like or clear fluids or cholesterol clefts and lined endothelial cells	Endothelial cells are positive for Factor VIII-related antigen and CD31
Littoral cell angioma	Presents almost always with splenomegaly	Anastomosing vascular channels lined by tall or flat endothelial cells with vesicular nuclei These channels may have a pseudopapillary aspect Plump exfoliated cells are often found within the lumina of the vascular channels	• Lesional cells are positive for ERG, factor VIII-related antigen, CD68, CD21 and lysozyme • The cells are negative for WT1, CD8 and CD34
Hemangioendothelioma	abdominal pain, palpable mass, or hypersplenism	The vascular and stromal components are both neoplastic with epithelioid or spindle shaped endothelial cells (or, a combination of these two patterns) with hyperchromatic nuclei. The epithelioid hemangioendothelioma variant does not have well formed vascular channels but rather, consists of individual cells with vacuoles.	Lining cells are positive for CD31, factor VIII—related antigen, and variably positive for CD34 and cytokeratin
Kaposi sarcoma (KS)	Four variants: classic KS, endemic (African) KS, iatrogenic (organ transplant-related) KS, and (AIDS)-related KS	Slit-like vascular channels with perivascular plump stromal spindle cells Promontory sign may be helpful Intracytoplasmic hyaline globules	• Lesional cells positive for CD31, CD34, and D2-40 • Antibody to HHV-8 (Human Herpes Virus 8) latent nuclear antigen 1 is highly specific
Angiosarcoma	Typical symptoms are fatigue, fever, weight loss and abdominal pain	Disorganized or irregular anastomosing vascular channel or solid masses lined by atypical endothelial cells with multilayering	CD31, CD34, factor VIII—related antigen, and CD68 are positive in tumour cells

Table 1

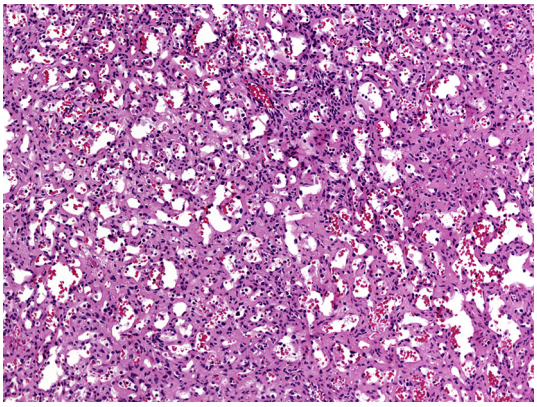

Figure 1 Haemangioma: Dilated blood vessels involving spleen. The lining endothelial cells lack cellular pleomorphism or hyperchromasia (Medium power).

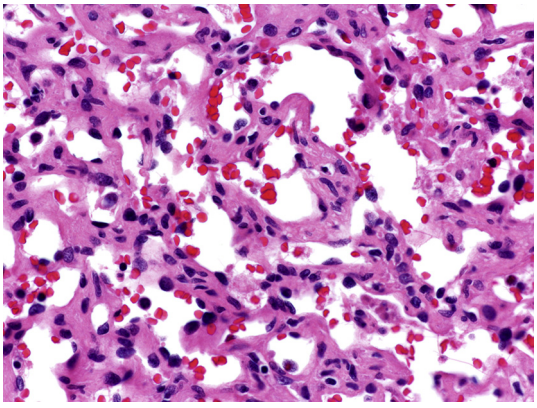

Figure 2 Haemangioma: Dilated blood vessels lined by bland endothelial cells (High power).