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ARTICLE

Tumours of the spleen†

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Abstract

The spleen has been considered a 'forgotten organ' even if it is included and well demonstrated on every CT and MRI of the abdomen. Tumours of the spleen are rare; however, radiologists need to be aware of the main tumoral features and patterns in order to try to distinguish between benign and malignant masses often discovered incidentally. The principal tumoral masses, benign (cysts, haemangiomas, litteral cell angioma, lymphangioma) and malignant (lymphoma, metastases haemagiosarcoma), are described.

Keywords: *Spleen; spleen imaging; spleen tumours.*

Introduction

The spleen has been considered a mysterious organ since classical times and nowadays it seems to be 'the forgotten organ', particularly among many radiologists. However, it is included and well demonstrated on every computed tomography (CT) scan and magnetic resonance image (MRI) of the abdomen and often on the lower part of chest CT scans. Although metastatic and primary tumours of the spleen are rare, radiologists need to be aware of the various tumoral features and patterns in order to recognise the benign or malignant masses, which often are found incidentally.

Splenic cysts

Many focal lesions may appear to be cystic on cross-sectional imaging. Cystic lesions can be classified as primary ('true') or secondary ('false'), based on the presence of a cellular or fibrous lining. Primary cysts can be divided into non-parasitic or parasitic (i.e. echinococcal). True non-parasitic cysts include congenital (i.e. epithelial) and neoplastic (lymphangioma, metastases, haemangioma) cysts. False cysts may develop secondary to trauma, haemorrhage, infarction-degeneration and inflammation.

Congenital or epithelial splenic cysts

Congenital or epithelial splenic cysts comprise approximately 25% of true cysts of the spleen. They are mainly seen in children and young adults and are usually solitary, but can be multiple. Although the exact mechanism of the aetiology, pathogenesis and development of congenital splenic cysts is unknown, proposed mechanisms include: involution of pleuripotent cells in the splenic parenchyma during development with subsequent squamous metaplasia; entrapment of peritoneal endothelial cells or coelomic mesothelium within the developing spleen, and invagination of the surface mesothelium or dilatation of normal lymph spaces. Microscopically, the wall is lined with columnar, cuboidal or squamous epithelium. They can be further subdivided as dermoid, mesothelial, and epidermoid. Dermoid cysts are extremely rare with only a few cases reported; they contain skin adnexa and squamous epithelium.

Generally, congenital splenic cysts are asymptomatic and the prognosis is good. Occasionally, congenital cysts may become symptomatic because of enlargement which may be secondary to trauma, haemorrhage from the

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cyst wall, an increase in the osmolality of the cystic fluid, or the presence of stomata in the cyst wall. Associated complications include infection, rupture and haemorrhage. The therapy of choice for a symptomatic splenic cyst is an interventional procedure such as partial or total splenectomy.

Typically on ultrasound a splenic cyst appears as a round, homogeneous, anechoic lesion with a smooth thin wall. Thin septations, an irregular cyst wall, or a mixed pattern of echogenicity from internal debris or haemorrhage and peripheral brightly echogenic foci with distal shadowing due to cyst wall calcifications may contribute to a more complex picture. An epidermoid cyst has a complex pattern with irregularity and thickening of the posterior wall because of epithelial peripheral trabeculation, and internal echoes from blood clots.

On CT, splenic cysts are typically spherical, well-defined lesions with an attenuation value near water and a thin or imperceptible wall which demonstrates no enhancement after an injection of contrast medium.

On MRI, the cyst is hypointense on T1-weighted images and strongly hyperintense on T2-weighted images, with a signal intensity equal to that of water with no enhancement after the injection of contrast medium. However, depending on the composition of the cystic fluid, the signal intensity on T1-weighted images may be increased (haemorrhagic cyst), while the signal intensity on T2-weighted images remains high.

False(pseudo) cyst

False(pseudo) cysts account for approximately 75% of the non-parasitic cysts of the spleen. They are secondary to trauma, infarction or infection. Trauma is the most probable aetiologic factor. The majority of these cysts are solitary and asymptomatic. Microscopically, the wall of these cysts is composed of dense, often calcified fibrous tissue with no epithelial lining. They contain a mixture of blood and necrotic debris. It is often impossible to distinguish radiologically between primary and secondary cysts; the clinical presentation and patient history may help to narrow the differential diagnoses.

Hydatid cyst

Hydatid cyst is due to a flatworm infection (echinococcosis) in which humans are an accidental intermediate host. It is the only parasitic cyst of the spleen. Splenic hydatid is extremely rare even in endemic regions with an incidence of 0.5%–4%. The cysts may or may not be calcified. Ultrasound and CT can establish the diagnosis definitively and daughter cysts, the presence of which is specific for echinococcal infection, can be identified.

On ultrasound, splenic hydatid cysts may be anechoic or of mixed echogenicity due to intra-cystic components (scolices). Separation of the membranes of the cyst produces the ‘waterlily sign’, whilst daughter cysts give the characteristic appearance of a ‘cyst within a cyst’.

On CT, the cyst is a round, low-attenuation cystic mass which may be unilocular or multilocular, often with discontinuous rim calcifications, and demonstrating no enhancement after intravenous contrast material administration.

Benign tumours

Most common primary tumours of the spleen are benign and originate from the vascular endothelium and include haemangioma, hamartoma, littoral cell angioma, lymphangioma, haemangioendothelioma, haemangiopericytoma. Non-vascular tumours comprise the inflammatory pseudotumour, fibroma and lipoma [1–6].

Haemangiomas

Haemangiomas are the most common benign tumour, found incidentally as they are usually asymptomatic. Most of them are less than 2 cm in diameter, but if the lesion is large there is a risk of spontaneous rupture with haemorrhage. They are mostly solitary but may be multiple or associated with haemangiomas of other sites (angiomatosis).

Two principal patterns are known: the cavernous variety (the most frequent) and capillary haemangioma. At histology, cavernous haemangioma consists of dilated vascular spaces filled with red cells and the capillary haemangioma is made up of small vascular spaces with a thin wall of capillary type [1].

The ultrasound pattern depends on the macroscopic type of haemangioma. Capillary haemangioma appears as a hyperechoic nodule whereas cavernous haemangioma is seen as a heterogeneous hypoechoic mass, sometimes with calcifications or multiple cystic areas.

On CT, capillary haemangiomas appear as small, well-marginated homogeneous iso- or hypodense masses with homogeneous contrast enhancement. Cavernous haemangiomas, usually of larger dimensions, appear more or less cystic with occasional iso- or hypodense areas and after the injection of contrast medium, they demonstrate early peripheral nodular enhancement with progressive fill-in and are homogeneous on delayed images, although they do not exhibit this typical enhancement pattern. Calcifications if present may be either peripheral and curvilinear or scattered centrally.

On T2-weighted MRI, haemangiomas exhibit a homogeneous hyperintense signal and a hypointense signal on unenhanced T1-weighted images. On dynamic enhanced T1-weighted images their morphologic behaviour is similar to that seen at CT and to that of hepatic haemangioma, with peripheral nodular enhancement and delayed persistent enhancement.

Hamartoma

Hamartoma is a rare benign tumour of the spleen, with an autopsy incidence of 0.13%. Approximately one-sixth of hamartomas are found in children (<16 years). They are usually <3 cm in diameter, but can reach up to 18 cm in size. Histologically, hamartoma is composed exclusively of red pulp components, but may also contain cystic or necrotic components and small calcifications. Spontaneous rupture of a hamartoma, with acute abdominal pain in adults has been reported, but most patients with splenic hamartomas are asymptomatic and the lesion is discovered incidentally.

Ultrasound may show a solid mass, which is sometimes heterogeneous with multiple hyperechoic areas assumed to represent punctate calcifications or cystic components. Colour Doppler ultrasound demonstrates a hypervascular lesion containing multiple radial blood flow signals.

On CT, hamartomas appear as well-demarcated, solid, hypodense masses, although a hyperattenuating appearance due to haemosiderin deposition has been reported. It demonstrates inhomogeneous and moderate contrast enhancement[2].

MRI may demonstrate a well-defined homogeneous mass which is isointense on T1-weighted images and slightly hyperintense on T2-weighted images. Dynamic enhanced T1-weighted images depict diffuse heterogeneous enhancement early after injection of contrast medium and more uniform enhancement is seen on delayed images.

Littoral cell angioma

Littoral cell angioma (LCA) represents a distinct new clinico-pathological entity of a very rare benign tumour of the spleen and develops from the lining cells of the red-pulp sinuses, the so-called ‘littoral cells’, giving rise to littoral cell angioma, first described in 1991. Although the tumour is considered as benign with no malignant histological features and has a benign clinical course, literature reviews reveal many other cases of littoral cell tumours with disseminated disease that may be examples of littoral cell haemangioendothelioma. Most patients are diagnosed in their 50s. They present with anaemia, pyrexia of unknown origin and a variable degree of splenomegaly leading to hypersplenism. However, a significant number of cases are asymptomatic and discovered incidentally post splenectomy[3].

Histologically, lesions are always situated within the red-pulp of the spleen, are of variable size and commonly multinodular. They are composed of anastomosing vascular channels with irregular lumina featuring cyst-like spaces and lined by tall endothelial cells. Histological description reflects the radiologic appearance of this tumour with splenomegaly and multiple round lesions of similar appearance and size diffusely distributed in the splenic parenchyma[4].

Ultrasound depicts lobulated spenomegaly with heterogeneous echogenicity or multiple nodules whose echogenicity is very close to that of the normal splenic parenchyma.

On CT, most commonly littoral cell angioma appears as multiple hypoattenuating masses in an enlarged spleen which on histopathologic examination, represent blood-filled vascular channels. After injection of contrast medium, minimal delayed enhancement is seen. In contrast to typical haemangioma, the internal morphology of LCA is inhomogeneous, and lesion distribution is also diffuse.

On MRI, lesions are inhomogeneously hyperintense on T2-weighted MR images, with signal similar to that of haemangiomas and slightly hypointense on unenhanced T1-weighted images. Dynamic enhanced T1-weighted images depict delayed contrast enhancement, suggestive of a vascular lesion with contrast media pooling[5].

Lymphangioma

Lymphangioma is relatively uncommon and frequently presents as a solitary splenic nodule or as a part of systemic lymphangiomatosis in young patients; both types are grossly cystic in most cases, but a solid lymphangioma with sclerotic change and papillary endothelial proliferation are also described. Histologically it is classified into three subtypes: simple (capillary), cavernous and cystic. Cystic lymphangioma is the most frequent type and is characterised by a honeycomb of large and small thin-walled cysts containing lymph-like clear fluid. Because the lesions are cyst-like, their appearance on ultrasound, CT, and MRI is similar to and indistinguishable from that of cysts.

On ultrasound, multiple hypoechoic cysts of various sizes with hyperechoic septae, debris and calcifications may be detected. Splenic lymphangioma may be differentiated by colour Doppler ultrasound, as a fan-shaped distribution of intrasplenic vessels extending from hilus to periphery.

Inflammatory pseudotumour

Inflammatory pseudotumour (IPT) is a term used to describe a tumorous fibroinflammatory process mainly occurring in liver and spleen. Histologically it is characterised by spindle cell proliferation admixed with abundant inflammatory cells, mainly lymphocytes and plasma cells. IPT lesions have been known as plasma cell granulomas and pseudosarcomatous myofibroblastic proliferations. At present, the preferred term of ‘inflammatory myofibroblastic tumour’ (IMT) is used, because it has been shown that the proliferating spindle cells show a myofibroblastic phenotype in many IPT cases. Inflammatory pseudotumour and IMT are descriptive terms, and thus include several entities that arise from

different aetiologies. That is, some IPT are reparative, some are infection-associated (latent infection by the Epstein–Barr virus), and some are neoplastic. Most cases are reported from eastern Asia and although their biological behaviour has not been fully investigated these neoplastic IPT are, at least, capable of recurrence and metastasis. The clinical presentation is non-specific, and some patients are asymptomatic. The tumours range in size from 3.5 to 22 cm, with most more than 10 cm in diameter[6].

On ultrasound, these lesions are well-circumscribed, hypoechoic masses.

On CT, inflammatory pseudotumour appears as a hypodense mass with delayed enhancement and there may be a central scar without enhancement corresponding to collagen fibres around vessels.

On MRI, these lesions are hypointense on T1-weighted images and hyperintense on T2-weighted images and following contrast medium there is inhomogeneous delayed enhancement similar to that previously described for hamartomas.

The other tumours, such as haemangioendothelioma, haemangiopericytoma, fibroma and lipoma, are extremely rare.

Malignant tumours

Malignant tumours of the spleen are uncommon with primary malignancies extremely rare[7–12].

Lymphoma

Lymphoma is probably the most common splenic malignancy and is usually a manifestation of generalised lymphoma. Primary splenic lymphoma is rare. Most of the primary splenic lymphomas are non-Hodgkin lymphomas (marginal zone cell lymphoma). The most common finding is splenomegaly, but it may be absent in up to 30% of lymphoma patients.

Ultrasound may depict a solitary lesion or slightly ill-defined inhomogeneous hypoechoic lesions. Another pattern is a general diffuse inhomogeneity with minute hypoechoic lesions less than 1 cm in size.

Staging of lymphomas on CT can be limited as only 45%–70% of lymphomas show diffuse splenic infiltration or tumour foci less than 1 cm in diameter so that the diagnosis of lymphoma can sometimes only be made microscopically. The focal lesions with diameter from 1 to 10 cm are typically of low attenuation and rarely enhance so may be better demonstrated on post-contrast scans[7].

MRI findings are non-specific and similar to those of metastases from other primary tumours. Typically, lymphomas are hypointense or nearly isointense on T1-weighted images and hyperintense on T2-weighted images. Injection of contrast medium may improve detection of splenic lymphoma[8].

Metastases

Although the spleen is the most vascular organ in the body, it is an infrequent site for metastatic disease (3.4% of metastatic carcinoma). Explanations proposed for the relative paucity of splenic metastases have included: the sharp angle made by the splenic artery which makes it difficult for tumour emboli to enter the spleen; the rhythmic contractile nature of the spleen which squeezes out the tumour emboli; the absence of afferent lymphatics to carry metastatic tumour to the spleen; and antitumour activity due to a high concentration of lymphoid tissue in the spleen. Apart from these factors, the frequency of splenic metastases may have been underestimated as they are often asymptomatic and occur late in the disease. Splenic metastases are most commonly found in malignant melanoma, lung, breast or ovarian carcinomas.

On ultrasound, they can show various degrees of echogenicity, but are usually hypoechoic[9].

On CT, splenic metastases typically appear as hypodense lesions which may be solid or cystic and with inhomogeneous contrast enhancement indicating a mixture of vascularisation or necrosis.

On MRI, metastases are predominantly hypointense on T1-weighted images and hyperintense on T2-weighted images, with occasionally inhomogeneous contrast enhancement. MRI is more accurate for the detection of splenic metastases which are necrotic or haemorrhagic.

Haemangiosarcoma

Haemangiosarcoma is rare, accounting for only 1%–2% of all soft tissue sarcomas, and is highly aggressive with poor prognosis[10–12].

Ultrasound and CT may show splenomegaly with a poorly circumscribed, inhomogeneous mass of variable echogenicity on ultrasound and of mixed attenuation with poor contrast enhancement on CT. Occasionally intratumoral necrosis and subcapsular or extracapsular blood collection can be seen because haemangiosarcoma may bleed and cause spontaneous rupture of the spleen.

The MRI appearance depends on haemorrhagic areas that are often present, and which suggest how tumour growth occurred over a short period of time. MR dynamic images clearly depict heterogeneous enhancement within the tumour, which corresponds to the pathologic findings of solid parenchyma with necrosis.

Conclusion

Tumours of the spleen are rare with cysts, haemangiomas, benign and malignant lymphomas being the most frequent. CT and ultrasound are used as screening modal-

ities for the spleen with MRI helpful for characterisation of lesions.

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