Spleen

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27.1	Anatomy and Characteristics Specific to Development - 816
27.2	Imaging – 816
27.2.1	Indications for Imaging of the Spleen – 816
27.2.2	Imaging Techniques – 817
27.3	Trauma – 818
27.4	Focal Splenic Lesions – 818
27.4.1	Cystic Lesions – 818
27.4.2	Splenic Tumours – 818
27.5	Infectious Disease – 823
27.6	Vascular Diseases – 823
27.7	Further Splenic Diseases – 824

The spleen is the largest lymphatic organ of the human body. A healthy spleen has the function to keep the concentration of blood cells in balance. Spleen disorders can cause a clotting disturbance as seen in general bleeding diathesis with haemorrhage following minor traumas or prolonged bleeding, e.g., following small surgical invasion.

While diseases of primary splenic origin are rare, congenital anomalies, injuries, and systematic diseases can lead to diffuse or focal abnormalities. Various factors can cause enlargement of the spleen, such as an immunodeficiency or infection, particularly oncologic, disease. Patients with pronounced spleen enlargement suffer pain or pressure in the left upper abdomen that can spread to the shoulder.

27.1 Anatomy and Characteristics Specific to Development

The spleen is located in the upper left quadrant of the abdomen below the diaphragm and costal margin. It is in medial and dorsal relation with the anterior surface of the left kidney and adrenal gland as well as the tail of the pancreas. It is medial and ventral to the stomach and caudally extends to the left colonic flexure.

The size of a healthy spleen in adults measures 4–5×7–8×11–12 cm (thickness × width × length).

A **normal spleen** consists of red and white pulp. The red pulp indicates the strongly vascular structure of splenic tissue, whereas the white pulp consists of lymphoid tissue, which can be the origin of lymphatic diseases and tumours. A discordant width of sinusoid in the red pulp forms a trabecular enhancement in the arterial contrast phase in CT and MRI (**•** Fig. 27.1). The spleen appears homogeneous in postcontrast imaging only when the red and white pulp receive equal distribution of the contrast agent.

Developmental defects of the spleen like asplenia are often coupled with other complex dysfunctions (e.g., situs inversus, heart abnormalities). Polysplenia can be accompanied by other congenital abnormalities or appear following splenic rupture and implantation of splenic tissue in mesenterium.

In **functional asplenia**, the organ is anatomically present but malfunctioning most likely because of damage to the reticuloendothelial system (RES). This is seen when uptake of superparamagnetic contrast agent or radioactive nuclear medicine tracers fail to be present. Functional asplenia was first diagnosed years ago through the detection of a missing accumulation of Tc sulphur colloid particles in an otherwise anatomically normal spleen.

Accessory spleens are found in about 10% of the population and may be either singular or multiple. They are usually located in the region of the splenic hilum and are often in contact with the tail of the pancreas. They generally occur as result of fusion failure and can be confused with enlarged lymph nodes or a growth in the tail of the pancreas. Their diameter can be 1–10 cm. Accessory spleens contain normal splenic tissue and



Fig. 27.1a,b Contrast-enhanced CT, normal result of the spleen. a Trabecular enhancement of the spleen in the arterial phase. **b** In contrast, a homogenous appearance of the spleen in the equilibrium phase

therefore reveal the same signal values in all procedures as the organ itself (**I** Fig. 27.2).

27.2 Imaging

27.2.1 Indications for Imaging of the Spleen

Ultrasonography remains the most important modality for the majority of primary indications for imaging of the spleen. Apart from trauma, CT and MRI are first used when diagnosis remains indeterminate following sonography. MRI is additionally used for diagnostic confirmation of special problems, e.g., within the context of storage diseases.

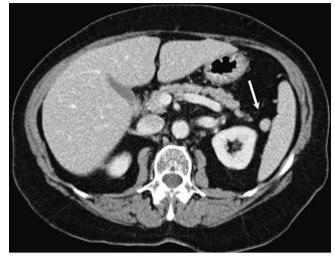


Fig. 27.2 Accessory spleen. Contrast-enhanced CT with evidence of a small accessory spleen above the splenic hilum and near the tail of the pancreas. Morphology and contrast agent behaviour correspond to the spleen

Indications for imaging of the spleen include:

- Determination of the size, form and location (esp. during therapy)
- Abdominal traumas
- Changes in structure, e.g., within the context of storage diseases, infections and systemic diseases
- Evidence of focal lesions: metastases, splenic tumours, infected lesions
- Residual condition following splenectomy

27.2.2 Imaging Techniques

Ultrasonography

Ultrasonography of the spleen uses a 3.5–5 MHz transducer and is preferably performed in the right lateral position. The cranial section of the spleen usually appears with deep inspiration under the costal margin. A normal spleen shows a homogeneous echo structure and, compared to its surrounding intestinal structures, appears mostly hypoechoic (a low echo) (Fig. 27.3).

Computed Tomography

Unenhanced and especially contrast-enhanced CT significantly improves the detectability of focal lesions. Unenhanced CT examination of the spleen is reasonable for the suspicion of haemorrhage or calcification. A normal spleen has an attenuation of 40–50 HU in CT, which is slightly lower than liver values.

Because contrast accumulation in the early phase is very inhomogeneous due to the ratios mentioned above, assessment of the spleen should always take place in the parenchymal phase. Note that this applies to examinations in the arterial phase, as performed in diagnostic CT of the pancreas or in the suspicion of hypervascular metastases in the liver.



Fig. 27.3a,b Sonography of the spleen. a Normal spleen with homogeneous echo structure with marked longitudinal diameter of 11.5 cm. b Splenomegaly in mononucleosis

Nuclear Medicine Imaging

Nuclear medicine procedures include either SPECT imaging with ^{99m}Tc-labelled tracers, e.g., colloid particles or heat-treated erythrocytes, or ¹⁸F-FDG PET and PET/CT. Primary indication for PET and PET/CT is rare, but because most imaging studies already include the spleen, it is important to take all correlating results into consideration.

Magnetic Resonance Imaging

MR imaging of the spleen uses T1W gradient echo sequences in axial and coronal planes with gadolinium-based contrast agent if required, just as for the parenchymatous upper abdominal organs. Moderately and strongly T2-weighted sequences are generally performed. A superparamagnetic iron oxide-based contrast agent can be used for the detection of small lesions. Following intravenous bolus administration and uptake of the superparamagnetic agent (e.g., Endorem), images demonstrate a significant decrease in signal intensity based on the high RES content of the spleen in T1W and T2W images.

In principle, the same pulse sequences are used for diagnostic imaging of the spleen as for imaging of the liver. In contrast to the liver, the spleen appears more hypointense in unenhanced T1W images and slightly hyperintense in unenhanced T2W images.

27.3 Trauma

Definition and Aetiology

Traumatic splenic rupture is mostly caused by blunt abdominal or penetrative trauma. Rupture may or may not include the splenic capsule following blunt abdominal trauma. Extreme splenomegaly, i.e., within the context of systematic diseases or infection, rarely leads to injury of the spleen. Clinical presentation usually includes pain in the area of the left shoulder as a result of phrenic nerve irritation. Whether the capsule ruptures within one hour or even days following the tear of the splenic tissue, it is referred to as a "double rupture." "Simple rupture" occurs immediately following trauma.

Imaging

In the suspicion of traumatic splenic rupture, sonography and contrast-enhanced CT are used to detect signs of intraperitoneal haemorrhage in the abdomen and to determine the tear of splenic tissue and cause of bleeding (Fig. 27.4). A crescentic collection of fluid usually appears under the splenic capsule if it is intact.

Ultrasonography shows splenic haematoma as hypoechoic or even anechoic with differentiating configurations. Small subcapsular haematomas in the cranial subdiaphragmatic area can be easily overseen. In CT, splenic haematomas in the acute setting appear hypodense. Older haematomas can appear isodense and can lack enhancement after injection of contrast agent.

MRI and other imaging techniques are not used in traumatic splenic rupture due to long examination times and high costs.

Ultrasonography and CT remain the imaging techniques of choice for trauma. The course of the patient's treatment depends on the determination of haemorrhage and the extent of damage to the splenic tissue.

27.4 Focal Splenic Lesions

Differential diagnosis of focal splenic lesions

- Simple, particularly infectious cysts
- Benign tumours: haemangioma, lymphangioma, hamartoma
- Primary tumours: lymphoma, angiosarcoma
- Secondary malignant tumours: metastases, especially in melanoma, bronchial carcinoma, and other tumours
- Abscesses (e.g., posttraumatic) like other infectious lesions, e.g., in sarcoidosis and Candida infection



Fig. 27.4 Splenic rupture in CT. Contrast-enhanced CT of the abdomen. Evidence of splenic rupture with extensive haematoma and intraperitoneal haemorrhage

27.4.1 Cystic Lesions

Definition, Epidemiology and Aetiology

Splenic cysts are relatively rare. They are commonly located around the inferior pole and subcapsular area. About one-fourth of all splenic cysts are congenital; acquired cysts can occur following trauma or infection. It is rare for secondary pseudocysts to grow in the spleen due to acute pancreatitis. Wall calcifications are frequent (in ca. 30–50% of all cases).

Imaging

Splenic cysts generally appear as well-defined lesions with water density.

In **ultrasonography** they appear hypoechoic with dorsal echo amplification. In **CT** cysts exhibit an attenuation value near water (<20 HE) and a thin wall. Increased peripheral vascularisation and contrast enhancement are not demonstrated. Proteinaceous content rises on account of internal bleeding and images therefore tend to exhibit hyperattenuation. Wall calcifications usually appear later in cysts caused by trauma. Parasitic cysts (e.g., echinococcus, etc.) often feature septations and wall calcifications.

In MRI cysts reveal a low attenuation signal in T1W images and a significantly high attenuation signal in T2W images. However, signal intensity can vary depending on proteinaceous content (• Fig. 27.5). Internal septations are low in intensity and reveal fibrous or reticular structures. In **nuclear medicine imaging**, cysts appear well localised and "cold."

27.4.2 Splenic Tumours

Benign Tumours

Haemangioma, hamartoma, and lymphangioma are the most common benign splenic tumours. Rare benign tumours include myxoma, chondroma, fibroma, and desmoids; their appearance is so rare that diagnosis frequently is made histologically following an operation.



Fig. 27.5a,b Splenic cysts. **a** T1W image following gadolinium DTPA administration. **b** T2W coronal image: smooth contours and significantly intense T2W image of lesions due to high fluid content equal to the liquor cerebrospinalis

Haemangioma

Definition and Pathogenesis Haemangiomas are intravascular tumours and are congenital in 75% of all cases. Splenic haemangiomas tend to appear singularly and can range in size from a few millimetres up to several centimetres. Isolated splenic haemangiomatosis is rare for this reason. Splenic haemangioma is predominately cavernous and rarely capillary, and arises from the sinus epithelium.

Because splenic haemangiomas grow slowly and are asymptomatic, therapy is generally unnecessary. Large haemangiomas are an exception, as spontaneous rupture may occur in up to 25% of all cases.



Fig. 27.6 Haemangioma. Correlating to typical contrast enhancement pattern of haemangioma, arterial phase CT exhibits nodular, circular enhancement

Imaging Ultrasonographic imaging reveals lesions with a hyperechoic appearance similar to haemangioma of the liver. Splenic haemangiomas are commonly detected as accidental findings following examination. They are smooth and rarely exhibit scars or central calcification.

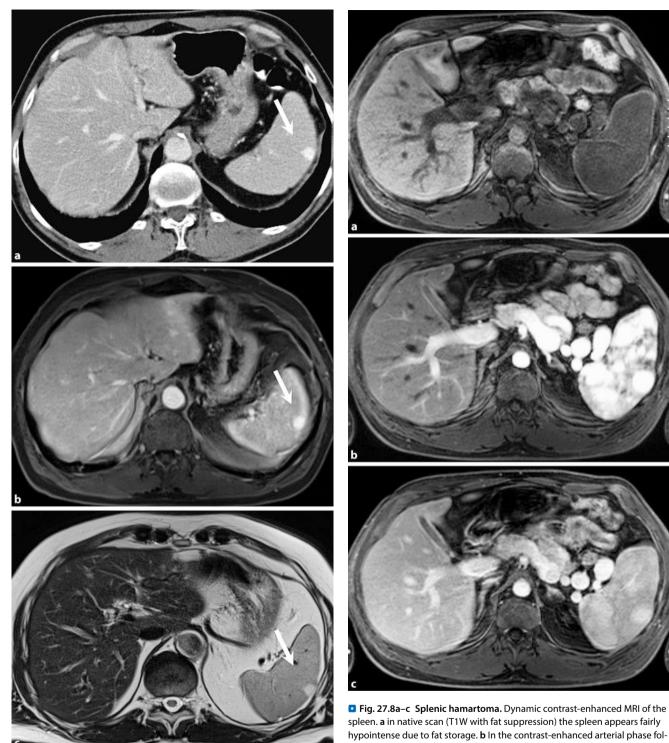
In **unenhanced CT imaging**, haemangiomas appear hypoattenuating and occasionally isoattenuating. Contrast administration usually produces peripheral enhancement in CT as well as in MRI (Fig. 27.6).

MR imaging appearance of splenic haemangioma shows increasing hyperintensity in T2-weighted images and centripetal progression of enhancement following intravenous bolus administration of a perfusion contrast agent like Gd-DTPA (Fig. 27.7). Nuclear scans with radioactive labelled erythrocytes are only used for haemangiomas difficult to diagnose (atypical, scarred haemangiomas). Uptake and storage of radioactive labelled erythrocytes reveal lesions as "hot spots."

Hamartoma

Definition and Pathogenesis Hamartoma (splenoma or nodular hyperplasia of the spleen) are seen as congenital, tumour-like neoplasms. They are usually singular and diagnosed as coincidental findings. Pathologically, hamartomas contain normal splenic tissue. Their size ranges from a few centimetres to large, whereas larger lesions may invade neighbouring organs.

Imaging Splenic hamartoma is a hypervascularised tumour, which tends to show strong contrast uptake from the centre outward. In addition to being rare, the evidence of fat (particularly in MRI) and calcification (preferably in CT) together in one finding can help making the diagnosis (**•** Fig. 27.8).



• Fig. 27.7a-c Splenic haemangioma of a patient in CT and MRI. a Homogeneous accumulation in the lesion appears in the late venous phase in CT. b A similar homogeneous enhancement appears following gadolinium DTPA administration in a T1-weighted fat-saturated sequence. c T2W image reveals a moderate hyperintense signal significantly lower than in splenic cysts

■ Fig. 27.8a-c Splenic hamartoma. Dynamic contrast-enhanced MRI of the spleen. a in native scan (T1W with fat suppression) the spleen appears fairly hypointense due to fat storage. b In the contrast-enhanced arterial phase following gadolinium injection, splenic hamartoma exhibits significant contrast uptake. c A homogeneous pattern of enhancement between lesions and normal splenic tissue can be found in the late phase (permission from Prof. Dr. Thomas VogI)

Lymphangioma

Definition and Pathogenesis Splenic lymphangioma occurs seldom and may be part of lymphangiomatosis, which affects other organs such as the liver, mediastinum, and skin. Histopathology of lymphangiomas reveals abnormal and insufficient excretory lymphatic tracks with small, consecutive calcifications. Because they are generally asymptomatic, surgical intervention via splenectomy is only necessary when secondary splenomegaly causes acute pain in the lower abdomen and compression of neighbouring organs.

Imaging Splenic lymphangioma is mostly subcapsular and appears in multiple masses. Sonography frequently reveals lesions as confluent cystic masses. In CT scans, lymphangiomas appear hypodense and show no significant contrast uptake. MR imaging also reveals cysts as confluent. They exhibit a hypointense signal in T1W images and a significantly hyperintense signal in T2W images.

The most important benign tumours are haemangioma, lymphangioma, and hamartoma. Their characteristic features make them relatively easy to classify following diagnostic imaging.

Malignant Tumours

Lymphoma

Definition and Epidemiology Lymphomas are the most common malignant tumours of the spleen. Splenic lesions occur in 20–30% of patients with Hodgkin's lymphoma and in 30–40% of patients with non-Hodgkin's lymphoma. Primary lymphoma is rare and has an incidence of 1%. Because the spleen in diffusely infiltrated in about 75% of all cases, focal lesions are often not evident. Enlargement of the spleen usually remains the only sign of evidence.

Diagnostics of splenomegaly

- Tumours: lymphoma, leukaemia
- Infections: mononucleosis, AIDS
- Vascular: portal hypertension
- Metabolic diseases, e.g., Gaucher's disease

Imaging Diffuse infiltration of the spleen appears inhomogeneous in unenhanced and contrast-enhanced CT. An inhomogeneous appearance is also common in MRI; T2W images often show the spleen as hypointense.

Following injection of a superparamagnetic contrast agent, significant uptake is not visual. In **small nodular infiltration** (<1 cm), nodular contrast enhancement is seen in CT and T1W MR images, and often with poorly defined margins. Although ultrasonography often underestimates the extent of micronodule infiltration, it continues to play an important role in staging and therapeutic progress.

Ultrasonography shows **larger lesions** (>1 cm) as hypoechoic. Lesion diameters in Hodgkin's lymphoma tend to be smaller than non-Hodgkin's lymphoma (\geq 3 cm). Diagnostic CT is also frequently used for lymph node staging.



Fig. 27.9a,b Non-Hodgkin's lymphoma. a Contrast-enhanced CT depicting coarse nodular lesions in region of spleen in a patient with non-Hodgkin's lymphoma. b¹⁸F-FDG PET/CT showing significant metabolic increase confluent throughout the whole spleen. Inhomogeneous accumulation in the liver (without diagnostic meaning) is also visible

PET and PET/CT are increasingly used on an increasing basis for diagnosis and staging as well as for the planning of therapy. Due to the lack of resolution, PET is limited for the diagnosis of single small splenic lesions compared to CT and MRI (**•** Fig. 27.9).

Symptoms and Therapy Clinical symptoms of splenic lymphoma are less determined by local findings as they are by the disease itself. The spectrum ranges from nonexistent (mostly in the initial stage) to common swelling of the lymph nodes as well as common B and secondary symptoms involving bone marrow. Although splenomegaly sometimes results in an unspecific feeling of pressure in the upper abdomen, it is seldom the leading symptom. Furthermore, the treatment of lymphomas is less focused on the enlargement of the spleen as on classifying the disease's stage on the whole; Ann Arbor staging recognises involvement of the spleen as stage IV.

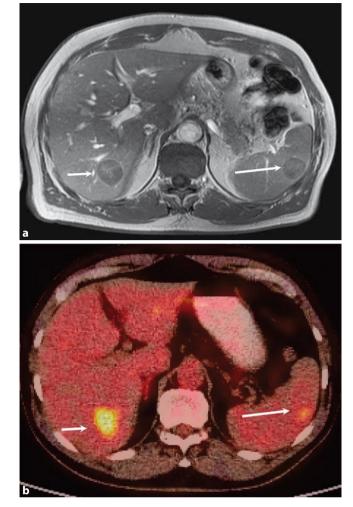


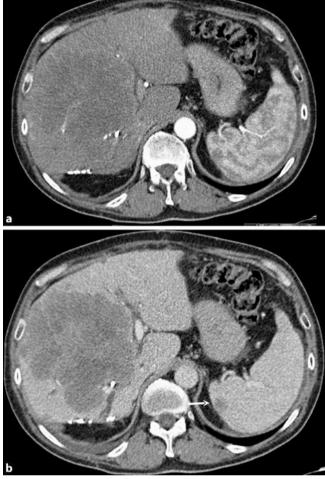
Fig. 27.10a,b Spleen and liver metastases. **a** Following IV injection of gadolinium DTPA, a hypointense metastasis appears in the liver and spleen with weak central enhancement in T1W images. **b** In ¹⁸F-FDG PET/CT there is significant tracer uptake in the liver metastasis and significantly lower uptake in the spleen

Primary Malignant Splenic Tumours

Though rare, the most common primary malignant tumour of the spleen is **angiosarcoma**, which occurs either de novo or due to exposure to Thorotrast, arsenic and vinyl chloride. All crossimaging techniques as well as angiography show angiosarcomas with increased vascularity. Such lesions are generally present in the liver when first diagnosed. **Haemangioendothelioma** is another rare tumour; it often appears in CT and MRI as a solid mass with necrotic areas. In both types of tumours, a splenic rupture can result in pronounced bleeding causing haemoperitoneum.

Metastases

Pathology Metastatic to in the spleen is usually present in advanced tumour diseases. The relatively late involvement of the spleen attributes to local immunologic mechanisms. Splenic metastases appear especially in mammary carcinoma, melanoma and bronchial carcinoma. Though seldom, they can also occur in



• Fig. 27.11a,b Colon carcinoma, advanced metastatic spread, contrastenhanced CT. a Typical inhomogeneous appearance of the spleen in the arterial contrast-enhanced phase, additional metastases are not clearly differentiable. Along the margins of the large mass in the liver are hyperdense formations from a previous partial resection. b In contrast to the arterial phase, a small marginal metastasis demarcates itself in the equilibrium phase on the lower splenic pole

ovarian, stomach, colon and prostate carcinomas. Metastasis of the spleen frequently reveals similar formations as in soft tissue organs, such as in the liver or lung.

Imaging Ultrasonography shows metastases as hypoechogenic. All cross-imaging techniques, however, tend to reveal reoccurring patterns of peripheral rings with elevated contrast enhancement and necrotic centres.

Hypervascular metastases already show contrast uptake in the arterial phase. In melanoma, metastases can also appear as hyperintense in T1W images due to high content of melanin. In most cases they also appear hypointense in T1W images.

With the help of modern nuclear medicine techniques, splenic metastases are detectable in PET and PET/CT as ¹⁸ F-FDG positive lesions (Fig. 27.10). Because splenic metastases do not occur until the advanced stage, diagnostic imaging and classification are often of minor relevance (Fig. 27.11).

27.5 Infectious Disease

Splenic abscesses are found mostly in patients with weakened immune systems, e.g., following chemotherapy. In patients with impaired immune system, abscesses occur following direct bacterial infection as well as tuberculosis, echinococcus infection, cytomegaly and varicella.

Patients with a weakened immune system are also susceptible to infections like *Candida albicans*, pneumocystis pneumonia, and *Cryptococcus neoformans*.

Infectious-specific abscesses can appear (In)

- Pancreatitis
- Perinephritic or subdiaphragmatic abscesses (per continuitatem)
- Metastatically as a result of sepsis
- Traumatically or postembolically as a super infection
- Immune deficiency
- Infectious diseases, e.g., echinococcus, tuberculosis, varicella, etc.

Imaging

All imaging modalities are able to detect bacterial abscesses with a diameter of only a few millimetres. The abscesses tend to exhibit typical radiologic patterns; fungal infections often appear very small and hypodense in CT. *Echinococcus granulosus* of the spleen shows similar changes in the diagnostic cross-imaging as in the liver. These cysts often present calcifications that are easily distinguishable in CT and sonography. T2-weighted MR images often reveal evidence of sepsis, which is valuable in the diagnostic process.

27.6 Vascular Diseases

Splenic Infarction

Definition, Aetiology, Clinic

Splenic infarction is mostly embolic or thromboembolic and occurs often in atrial fibrillation or due to endocarditis with leftventricular thrombus formation. Further causes are haemolytic anomalies or systematic lupus erythematosus as well as possible venous infarction in the splenic sinusoids in patients with thrombosis. While small infarctions are often asymptomatic, larger infarctions are often accompanied with pronounced abdominal pain and fever.



Fig. 27.12 State after splenic infarction with detection of calcifications in the upper pole region of the spleen

Imaging

Ultrasound shows splenic infarction in the early stage (<5 days) as inhomogeneous hypoechoic; the infarction area at this point is not very well defined. In CT infarctions appear as hypodense and partially inhomogeneous areas, and in T1W MRI as hypointense areas. They are mostly located in subcapsular area and present a triangular, round (although rare) or linear form. Haemorrhage in the acute phase can impress hyperdense in CT and especially hyperintense in T1W images.

In long term course, the infarction area may present a pseudocyst or secondary calcification (**•** Fig. 27.12). As the infarction area shrinks, a retraction of the spleen's surface becomes visible in advanced stages. Although smaller splenic infarctions can appear in sonographic images, CT and MRI provide clear evidence. Lack of contrast uptake is especially evident in advanced stages.

Splenic Vein Thrombosis

Pancreatitis is the principle cause of splenic vein thrombosis, although tumours of the pancreatic tail, retroperitoneal neoplasms, and traumas are also included.

Imaging. Doppler sonography, contrast-enhanced CT and MRI detect thrombosis of the splenic vein. Angiography is therefore seldom used in this case.

Splenic Artery Aneurysm

Spurious aneurysm can appear in trauma (see above) due to an effective shearing force (belt injury). It is necessary to differentiate trauma-related spurious aneurysm from congenital aneurysm of the splenic artery, which often appears together with other arterial aneurysms (coeliac trunk, hepatic arteries).

Imaging. Ultrasonography may detect splenic artery aneurysms as pulsating sacculations of the splenic artery depending on size. CT or MRI, on the other hand, are able to detect even the smallest aneurysms, where, due to physiological winding of the spleen (artery), partial sectioning may not be misinterpreted.

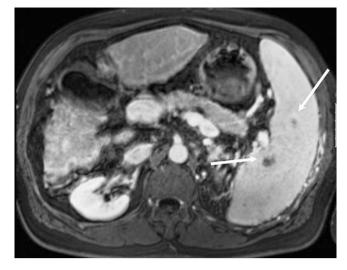


Fig. 27.13 Portal hypertension in hepatic cirrhosis, Gamna Gandy bodies. In the equilibrium phase of a T1 GRE sequence, areas reveal significant hypointense behaviour due to susceptibility artefacts of iron storage. Coarse nodular changes in the liver are also present

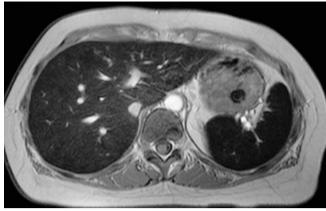


Fig. 27.14 Haemosiderosis. MRI of haemosiderosis specific to transfusion. Note the significant decrease of signal intensity in liver and spleen in the T1-GRE sequence

ing reveals iron accumulation in a similar fashion to secondary haemochromatosis.

Portal Hypertension

For definitions and aetiology, etc., see ► Sect. 24.4.

Imaging. Portal hypertension may be caused by elevated local pressure that leads to trabecular and perifollicular haemorrhage. CT reveals these haemorrhages as micronodular areas because of high iron content. In MRI, the so-called Gamna Gandy bodies appear especially when using T1W gradient echo sequences as well as a T2W images due to their susceptibility as varying large low signal lesions (**2** Fig. 27.13).

27.7 Further Splenic Diseases

Haemochromatosis

Primary haemochromatosis, caused by excess iron absorption, is typically characterised by increased iron deposition in the liver, pancreas, heart, etc., but not the spleen. All medical imaging techniques can detect haemochromatosis, in particular MRI, where normal signal intensities are presented in T1W and T2W images.

Secondary haemochromatosis with haemosiderosis, usually resulting from multiple blood transfusions, is characterised by an iron overload in reticuloendothelial tissue including the spleen. The spleen and liver appear significantly hyperdense in CT, depending on the extent of iron accumulation. MRI shows a significantly decrease in signal intensity in T1W and T2W images (Fig. 27.14).

Sickle Cell Anemia

All imaging techniques detect a greatly enlarged spleen in sickle cell anemia. Severe forms generally lead to a complete loss of splenic function. Multiple infarctions may be associated with relative splenic enlargement. Haemosiderosis with iron accumulation in the reticuloendotheliosis is a common complication of heterozygous sickle cell anemia in adults. In this stage, imag-

S Conclusion

- All imaging techniques are able to detect the spleen.
- The spleen commonly appears inhomogeneous in diffuse enlargement. The specific reasons of splenic involvement can only be assessed taking into account the complete clinical presentation.
- Ultrasonography and CT are preferred for acute confirmation in trauma, as they are significantly less time consuming than MRI.
- Compared to CT and ultrasonography, MRI generally demonstrates higher value in the diagnosis of focal lesions. Superparamagnetic iron-based contrast agents may be used for the detection of smaller lesions. Finally, MRI proves particularly superior in cases of storage diseases with iron accumulation due to sensitivity to signal changes.