Learning objectives

The focal mass in liver of child is not uncommon problem in the daily radiologists’ work. Our aims in this exhibit are to:-

• Provide a list of the common and uncommon liver masses in pediatric age group.
• Recognize the imaging features of each of uncommon liver mass in children.
• Describe how to distinguish liver tumors from other tumors on the basis of clinical and imaging data.

Background

The focal hepatic lesion is not uncommon problem in pediatrics radiology practice. Hepatic masses constitute about 5-6% of all intra-abdominal masses in children. The majority of liver tumors in children are malignant. Malignant tumors of liver constitute the third most common intraabdominal malignancy in the pediatric age group after Wilms’ tumor and neuroblastoma. Only about 1/3 of the liver tumors are benign.

Metastasis and hepatoblastoma are the most common and well known hepatic lesions. Yet a long list of other lesions presented as a hepatic mass.

A differential diagnosis of liver tumors in children can be obtained based on the age of the child, clinical information (in particular AFP) and imaging characteristics.

Reorganization of the radiological criteria of these lesions is essential to early diagnose and proper mange such lesions in children

Imaging modalities

The palpable mass is the commonest presentation of the liver tumors in pediatrics. Other presenting symptoms include pain, anorexia, jaundice, paraneoplastic syndromes, hemorrhage, or congestive heart failure.

By clinical examination, the abdominal mass usually obvious, yet the organ of origin is often not clear without imaging. Pediatricians and surgeons began to order more imaging studies because advances in imaging technology improved the diagnosis and management of disease. Because surgical resection remains the mainstay of treatment for many of these lesions, detailed depiction of the extent of the mass and relationship to hepatic anatomy is essential. Multiple imaging modalities are used to characterize the hepatic masses such as ultrasound, CT scan, MR imaging, angiography, and radionuclide techniques.
In imaging children, the risks of radiation exposure and sedation must be weighed, and the diagnostic approach should be tailored to the differential diagnosis appropriate for that child based on age and clinical data.

**Ultrasound** is the primary imaging modality in the radiological assessment of the abdominal mass especially in the children. It is an excellent screening study. It is widely available relatively cheap modalities. The lack of hazards of ionizing radiation and need for sedation are great merits in pediatrics imaging domain; Ultrasound accurately excludes a mass when it is not present and identifies the organ of origin when a mass is present. Identifying the organ of origin helps determine the remainder of the child's imaging work-up. US differentiate reliably solid and cystic lesions, and assess vascular flow also used successfully to guide the biopsy for tissue pathological diagnosis. Intraoperative US may also be used to guide resection and define vascular anatomy.

Usually additional imaging is obtained with CT scan or MR imaging.

Although using ionizing radiation, **CT** has merits of being fast requires less or no anesthesia. So CT has always played a major role in the imaging of the liver. But at the same time pediatric patients present unique technical challenges for CT. Children are not simply small adults, and CT scan principles drawn from experience with adults can not accurately be extrapolated to the pediatric population. The methods of CT examination should be adjusted.

With fast modern multidetector computed topmographic (CT) scanners, sedation is not required and the risks of radiation can be significantly reduced by using dose reduction techniques.

Recently the MRI is increasingly used to assess the pediatric liver masses, having a merit of no ionizing radiation and better tissue characterization yet require anesthesia as it has a long scan time.

Both CT and MRI are used in hepatic lesion detection and characterization as well as the preoperative mapping., angiographic studies and volumetric assay are usually used in this domain.

Whether CT scan or MR imaging is the modality of choice for definitive imaging of liver masses is a controversial issue. The choice is usually based on institutional experience and modality availability.

**Findings and procedure details**

Pediatrics hepatic tumors are classified into benign and malignant categories. Malignant hepatic neoplasm are twice as frequent as benign neoplasms.
Malignant liver tumors account for slightly > 1% of all pediatric malignancies.

Two thirds of liver tumors in children are malignant. The most common neoplasm involving the liver in children like the adult is metastatic disease, usually from neuroblastoma, Wilms tumor, or lymphoma.

The primary pediatric liver cancer includes both lesions unique to the pediatric age group and others that are more common in adults. The assessment of the child with liver mass should include the age of the patient, laboratory findings, and specific imaging features.

Hepatoblastoma accounts for two thirds of liver tumors in children. Other liver malignancies in children include sarcomas, germ cell tumors, and rhabdoid tumors, as well as the more familiar hepatocellular carcinoma.

1. **Hepatoblastoma**

Hepatoblastoma is the most common primary malignant tumor of the liver in children. For poorly understood reasons, hepatoblastoma occurs in males significantly more frequently than it does in females, male-to-female predominance of 1.5:1 to 2:1.

The age distribution of disease in childhood is notable, with 68% of cases manifesting in the first 2 years of life and 90% seen in patients younger than 5 years of age. Four percent of cases are congenital. Less frequently, hepatoblastomas may also occur in older children, up to 15 years of age.

Hepatoblastoma has been associated with several syndromes, including Beckwith-Wiedemann syndrome, Gardner syndrome, familial adenomatous polyposis, type 1A glycogen storage disease, and trisomy 18. Approximately 5% of cases occur in conjunction with other congenital anomalies, commonly of the genitourinary and gastrointestinal systems.

Most commonly, these tumors present in the right lobe of the liver.

Hepatoblastomas are composed of cells resembling the embryonic liver, hence the classification as an embryonal tumor. Indeed, the cells comprising hepatoblastoma mark similarly to hepatic stem cells, defined as pluripotent hepatoblasts capable of differentiating into hepatocytes or cholangiocytes.

Clinically, the hepatoblastoma present with enlarging abdomen and nonspecific symptoms including anorexia and weight loss. 5% of patients present with jaundice. Metastatic disease most frequently involves the lungs, with pulmonary metastases seen in 10%-20% of cases. Other common sites of metastasis include the bone, brain, lymph nodes, eye, and ovary.
Alfa feto protein (AFP) assay is useful laboratory marker for hepatoblastoma is important not only in the diagnspois but also serve as a marker to monitor therapy and detect recurrence. About 90% of patients with hepatoblastoma show abnormal elevation of AFP level. Normally the serum AFP levels are elevated at birth, measuring 25,000-50,000 ng/mL, and do not reach the normal adult level of less than 25 ng/mL until 6 months of age. The tumor hepatoblasts also may secrete human chorionic gonadotropin, which may manifest in features of precocious puberty in boys.

The imaging characteristics of hepatoblastoma reflect its gross pathologic appearance and histologic composition, as the epithelial subtype usually shows a homogeneous appearance, while mixed epithelial and mesenchymal tumors appear more heterogeneous due to osteoid, cartilaginous, and fibrous components.

**Plain Abdominal radiographs** usually demonstrate enlarged liver shadow. Coarse or dense calcifications may be noted due to the presence of osteoid material in mixed tumors.

**US** :- Heptoblastomas are most often hyperechoic relative to adjacent liver. Again the epithelial subtype may appear homogeneous and the mixed tumors are heterogeneous and often contain echogenic shadowing calcifications and anechoic foci representing hemorrhage and necrosis. Hypoechoic fibrotic septa may be observed.

**CT** :- Unenhanced CT typically shows a relatively well-defined, heterogeneous mass, slightly hypodense compared with liver tissue. Speckled or amorphous calcification is seen in more than 50% of lesions. Area fat density may also seen. On contrast-enhanced CT, the tumor reveals a heterogeneous enhancement. The tumor enhances during the hepatic arterial phase of dynamic contrast enhanced CT and becomes hypoattenuating in the portal venous phase of enhancement.

The tumor thrombus can invade the portal vein, spread along inferior vena cava (IVC) and encroach in the lumen of right atrium.

Metastasis may be seen in lymph nodes and lung parenchyma, rarely in the bones and brain.

**MRI** :- epithelial hepatoblastomas are homogeneously slightly hypointense on T1-weighted images and hyperintense on T2-weighted images relative to adjacent liver parenchyma. Mixed tumors demonstrate more heterogeneous signal intensity characteristics. Fibrotic septa are hypointense on both T1- and T2-weighted images and enhance after intravenous administration of gadolinium contrast material. Calcifications, which are more common in mixed tumors, are not well seen at MR imaging. Areas of hemorrhage may appear hyperintense on T1-weighted images, and vascular invasion is best demonstrated with gradient-echo sequences. MR angiography is useful to evaluate the hepatic vasculature preoperatively.
1. 2. HEPATOCELLULAR CARCINOMA

Hepatocellular carcinoma, malignant tumor of cells of hepatocyte differentiation, the second most common primary malignant liver tumor in pediatrics. HCC has a median age of 12 years, with a range of 5 to 15 years, and rare under 5 years, so the hepatocellular carcinoma is the most common hepatic malignancy of adolescents. Boys are more frequently affected, with a male-to-female ratio of 1.8:1 to 2.2:1.

Often, hepatocellular carcinoma is associated with known hepatic viral infection or cirrhosis, and while it can take decades for malignancy to develop, occasionally cases are seen in very young children. In regions without endemic hepatitis B infection, preexisting liver disease is found in about 50% of patients. Predisposing conditions include biliary atresia, familial cholestatic jaundice, glycogen storage disease type 1, antitrypsin deficiency, Wilson disease, hereditary tyrosinemia, and cirrhosis in addition to hepatitis B or C infection.

Occasionally, malignant tumors in children are seen with features of both hepatocellular carcinoma and hepatoblastoma. These tumors are more common in children with a diagnosis at later ages than that typical of hepatoblastoma.

Clinically: the children usually present with an abdominal mass and may also have abdominal pain, weight loss, fever, or anorexia (14). Serum AFP levels are markedly elevated in 70% of patients.

Imaging Features

The imaging features of the pediatric HCC is similar to that of the adult one

US appearance of HCC is variable. Small HCC is mostly hypoechoic to normal liver yet it may be isoechoic or hyperechoic. Large HCC usually appears more heterogeneous with hyperechoic areas representing fat or acute hemorrhage and anechoic areas due to necrosis or old hemorrhage. A thin hypoechoic halo may be noted in tumors with capsules. Infiltrative HCC may appear as a diffuse disruption of the normal liver echotexture.

The HCC shows high-velocity arterial flow on Doppler evaluation, it is also useful in detecting tumor thrombus in the portal or hepatic veins or inferior vena cava.

CT :- HCC may be homogeneous or heterogeneous, focal or infiltrative, solitary or multifocal, well- or ill-defined. On non enhanced scan, HCCs are typically isodense or slightly hypodense compared with liver parenchyma. A mosaic appearance is more commonly seen in larger lesions owing to components of hemorrhage, necrosis, fat, and calcification.
Dynamic contrast CT scan, HCC is predominantly supplied by the hepatic artery so it usually shows an early arterial contrast enhancement and a rapid wash-out on enhanced CT. (Fig.3). capsular enhancement in the delayed scans may be also detected. Vascular invasion of portal veins, hepatic veins, hepatic arteries and inferior vena cava may be seen. Rarely, tumor rupture may be seen with intra- or extrahepatic hemorrhage.

The diagnosis of underlying cirrhosis may help in the differential diagnosis, but is rare in children. Diffuse involvement of the liver leads to a diffuse hypodense liver on CT. HCCs metastasizes to lung, bone, skin and brain.

**MRI:** HCC typically is hyperintense relative to normal liver on T2WI with variable in signal intensity on T1wi. In larger lesions, the signal intensity is often heterogeneous, resulting in a mosaic pattern, which corresponds to foci of hemorrhage, fat, necrosis, copper, and calcification.

The enhancement pattern is the same as in the dynamic CT study.

Vascular invasion appears as lack of a signal void on spin-echo images and as an arterial enhancing mass with a delayed filling defect on dynamic gadolinium-enhanced images. Extracapsular extension of the tumor and satellite nodules may be seen.

### 1. **3. Undifferentiated (Embryonal) Sarcoma**

Undifferentiated embryonal sarcoma (UES) is the third most common hepatic malignancy, after hepatoblastoma and hepatocellular carcinoma. It is an aggressive tumor of mesenchymal origin, previously known as malignant mesenchymoma, mesenchymal sarcoma, embryonal sarcoma, or primary sarcoma of the liver. The term *undifferentiated (embryonal) sarcoma* was proposed by Stocker and Ishak in 1978 due to the primitive appearance of cells and the lack of histologic differentiation at light microscopic examination.

UES are usually diagnosed in children 6-10 years of age, but the tumor also occurs in younger children and adults. There is a slight male predominance.

**Clinically:** it usually presented with abdominal mass and pain. The serum AFP assay is consistently normal. Advanced cases metastasize to the lungs, pleura, and peritoneum. Vascular ( IVC ) invasion is rare.

**Imaging Features:** The solid appearance on US while cystic on CT and MRI is a unique feature help in diagnose this tumor. These appearance may attributed to the high water content of the prominent myxoid stroma.

**Plain radiography:** UES is seen as a large, usually with no calcification.
US: - UES usually appears solid, this appearance correlates well with the pathologic findings (83% of tumor volume is solid), iso- to hyperechoic relative to normal liver with small anechoic spaces. These anechoic areas correlate with foci of necrosis, old hemorrhage, and cystic degeneration at macroscopic examination (Fig. 4).

CT: - reveals predominantly cystic attenuation (88% of tumor volume) with solid foci, usually at the periphery or forming septa of variable thickness. This appearance correlates with myxoid stroma and pseudocapsule at gross examination. Central foci of high attenuation representing acute hemorrhage may also be seen. Calcifications are uncommon. Contrast enhanced CT shows predominantly peripheral enhancement is noted on delayed images.

MRI: - UES exhibit CSF like signal on T1- and T2wi. The capsule appears as a hypointense rim on T1- and T2wi. Hemorrhagic foci (T1 hyperintensity and T2 hypointensity) are often seen. Fluid levels may be identified. Internal debris and septa are well shown on T2wi. MR imaging is superior to CT for determination of resectability and evaluation for involvement of venous structures, the biliary tree, and adjacent lymph nodes.

1. Embryonal Rhabdomyosarcoma

Rhabdomyosarcoma is the most common soft-tissue sarcoma in children, it is a highly aggressive tumor and can occur anywhere in the body, the rhabdomyosarcoma of the biliary tree is rare and represents 1% of pediatrics liver tumors. It occurs almost exclusively in children. It is usually diagnosed under the age of 5 years.

Clinically: it present by jaundice and may be accompanied by abdominal distention, fever, hepatomegaly, or nausea and vomiting clinically it usually overlapped with much more common diagnosis of viral hepatitis. The level of levels of predominantly conjugated bilirubin and alkaline phosphatase is elevated and AFP level is normal. Metastatic disease is present at diagnosis in as many as 30% of cases.

The radiological features of biliary rhabdomyosarcoma are quite variable. Biliary duct dilatation with mass seen within ducts. When the rhabdomyosarcoma arises from the common bile duct, it often located in or near the porta hepatis, less common it arises from the intrahepatic biliary ducts, it appears as hepatic focal lesion.

US: - the tumor may appear as a solitary heterogeneous hypoechoic mass or multiple hypoechoic nodules separated by septa. Portal vein displacement without thrombosis is common.

CT: - homo- or heterogeneous hypo- or hyperattenuating mass, which may have prominent fluid-attenuation components. In the post contrast study, the pattern
of enhancement is highly variable, ranging from intense heterogeneous globular enhancement to none. (Fig. 5).

**MRI:** - T1 hypointense and T2 hyperintense or predominantly fluid-intensity mass in the common duct or in biliary radicals or a heterogeneous intrahepatic mass with large fluid-intensity areas.

**Percutaneous or operative cholangiography** frequently reveals a large, irregular or polypoid intraluminal mass

**Gallium uptake** has been reported and may be useful in localizing metastatic disease.

1. 5. **Hematological malignancies (Lymphoma, leukemia)**

More than 50% of the lymphoma (Hodgkin disease and non-Hodgkin lymphoma (NHL)) involves the liver.

Primary hepatic lymphoma is rare, while the secondary lymphomatous involvement of the liver is more common in both pediatric and adult patients. The immunosuppressed states are considered as risk factor for developing primary hepatic lymphoma.

**CT:** The lymphoma can affect liver either in focal or diffuse unifiltrative pattern:

- **Focal type** :- primary hepatic lymphoma may appear isodense to hypodense compared with normal hepatic parenchyma while the secondary hepatic lymphoma usually appears as multiple hypodense lobulated masses. (Fig. 7).

**The infiltrative type** may present as a diffusely hypodense liver. (Fig. 8).

Hepatic lymphoma does not show substantial contrast enhancement.

**On MRI,** lymphoma is hypointense on T1wi images and hyperintense on T2-wi and usually shows only minimal contrast enhancement.

1. 6. **Atypical teratoid/rhabdoid AT/RTs tumor**

Atypical teratoid/rhabdoid AT/RTs tumor is an uncommon aggressive pediatric neoplasm of uncertain origin. It was first identified in the kidney of infants and children and was described in 1978 as rhabdomyosarcomatoid variant of Wilms’ tumor. It was not recognized as a distinct entity until the 80s. It commonly occurs in central nervous system and kidney. Recently some reports about occurrence in other anatomical location rather than the kidney and CNS. The first evidence in literature of a liver tumour with rhabdoid features was in 1982 by Gonzalez-Crussi et al. The ATRT occurs at the infancy and young
children, usually presented by palpable masses. US is usually the first screening tool followed by CT. Transabdominal ultrasound revealed a large heterogeneous mass with intratumoral arterial vascularization. The large heterogeneous solid mass is the common radiological presentation. The lymphatic deposits are rare. (Fig. 9).

1. **7. Metastatic Disease**

The liver is one of the commonest sites for distant spread of neoplasms. The secondary hepatic tumor is much more common than primary hepatic malignancy in the pediatric population. The most common primary tumors that metastasize to the liver are neuroblastoma and Wilms tumor.

**CT:** - metastases usually produce multiple focal hypodense or hyperdense masses of varying size that are distributed randomly throughout the liver parenchyma. (Fig. 10&11).

**MRI:** - hepatic metastases are generally hypointense on T1wi. Variable signal on T2wiusually ranging from intermediate to high signal intensity, depending on the primary tumor. The enhancement pattern also depends on the primary tumor.

1. **II. Benign pediatrics liver tumors :**

Although benign hepatic neoplasm represent only a small percentage of solid liver tumors that occur in children. One third of primary liver tumours in children are benign.

1. **Infantile haemangioendothelioma**

Infantile haemangioendothelioma(IHE) or infantile hepatic hemangioma is the commonest pediatric benign liver tumor. It is a vascular tumour. The natural history is rapid proliferation followed by involution. Ninety per cent of lesions present in the first six months of life, one-third within the first month. They are rarely discovered after 1 year of age, and biopsy is indicated for older patients to exclude malignant tumors. There is a slight female . There is an increased prevalence in patients with hemihypertrophy and Beckwith-Wiedemann syndrome.

About one-half of cases occur as solitary masses and one-half are multifocal.

Clinically: IHE usually presented with presentation may be with a liver mass or CCF secondary to large arteriovenous shunts or consumptive coagulopath (Kassabach-Merritt syndrome).

Severe hypothyroidism may be caused by high levels of type 3 iodothyronine deiodinase activity produced by the tumor; the hypothyroidism may lead to cardiac dysfunction and mental retardation. Rarely, patients may present acutely with hemoperitoneum due to tumor rupture.
Laboratory tests can show anaemia and mild hyperbilirubinaemia but AFP is not raised.

**Imaging features**

Depend on whether the lesions are focal, multifocal, or diffuse and reflect their vascular nature.

- **Multifocal lesions** are small and uniform in appearance (Fig.12).

- **Large focal lesions** often demonstrate findings related to central hemorrhage, necrosis, fibrosis, and calcification.

- **In diffuse disease**, the liver is massively enlarged and replaced by multiple large masses, causing mass effect on adjacent organs and compression of the inferior vena cava.

Typically, evidence of high flow is apparent, as manifest by enlargement of the hepatic arteries and veins and possibly tapering of the abdominal aorta below the origin of the celiac axis.

**Enhancement pattern** is diagnostic. It shows intense peripheral nodular enhancement on arterial phase followed by progressive centripetal infilling on portal venous and delayed imaging. In large lesions with central fibrosis and necrosis, complete central enhancement may not occur.

Speckled calcification is noted in 50%.

Owing to the risk of bleeding, biopsy of these masses is avoided, and the diagnosis is made on the basis of typical imaging findings and the demonstration of involution at follow-up.

Patients with multiple liver lesions should be evaluated with **chest radiography and brain imaging** to exclude additional lesions.

**At prenatal US**, polyhydramnios and a hypoechoic liver mass may be detected. Findings of fetal hydrops, including anasarca, ascites, pleural effusion, and cardiomegaly, should be sought because these have prognostic import.

**Angiography** was the mainstay of diagnosis in the past but has been largely supplanted by dynamic contrast-enhanced MR imaging and CT. Angiography is now reserved for patients with intractable complications from arteriovenous shunts in whom use of embolotherapy is contemplated. Arteriography frequently shows dilated tortuous feeding arteries including the hepatic arteries and adjacent systemic arteries. Early draining veins due to arteriovenous shunts are seen. Focal lesions may have large venous varices with anomalous draining veins.
2. Mesenchymal hamartoma

Mesenchymal hamartoma of the liver is the second most common benign liver mass in children after infantile hemangioendothelioma. It is most commonly discovered in children younger than 2 years of age, with nearly all lesions (95%) discovered by age 5 years. There is a slight male predominance (3:2).

Clinically: it is commonly presented by (MH) present with abdominal distension, abdominal pain or incidentally on antenatal scans. The abdominal enlargement is usually gradual, although distention can develop fairly rapidly due to fluid accumulation within the cysts. Liver function tests and AFP are normal.

Imaging Features

The mesenchymal hamartoma has a spectrum of imaging features from a predominantly cystic mass with thin or thick septa to a predominantly solid (stromal or mesenchymal) mass containing a few small cysts.

Mesenchymal hamartoma are usually solitary although occasionally multifocal.

US:- cystic lesions contain low level echoes with echogenic septae and echogenic solid components. Calcification is rare.

CT:- complex cystic mass with enhancement of the septa and solid components. Low-level echoes may be seen within the fluid, presumably reflecting gelatinous contents. Color Doppler imaging shows relatively little blood flow, which is limited to solid portions and septa (Fig. 13&14).

MRI :- again depend on the cystic versus mesenchymal components and protein content of the fluid. Solid portions are hypointense on T1 sequences but variable on T2 sequences due to fibrosis, while cystic components are hyperintense on T2 sequences and variable on T1 sequences depending on the protein content. Enhancement is limited to septa and solid components.

1. Hepatocellular Adenoma

Hepatocellular adenoma, or hepatic adenoma, is a rare benign hepatic neoplasm that associated with the use of steroids. The hepatic adenoma usually occurs in adult women with a mean age of 30 years. Pediatric patients mainly consist of girls over 10 years old. Hepatic adenoma have been reported in association with several diseases, particularly glycogen storage disease types I and III, and also galactosemia and familial diabetes mellitus. There is also an association with congenital and acquired abnormalities of the
hepatic vasculature, such as portal vein absence or occlusion, and other hypervascular hepatic neoplasms, including adult hemangioma and FNH

Clinically, the main clinical concern is intratumoral hemorrhage, which occurs in 10% of patients, or, less common, rupture with intraperitoneal hemorrhage and hypovolemic shock. More commonly, adenoma is asymptomatic or present with an abdominal mass. Chronic and acute abdominal pain are other reported symptoms. Results of liver function tests are usually normal with no elevation of AFP level.

Imaging Features

The imaging appearance of hepatic adenoma depending on its pathologic composition. The lesion without hemorrhage are homogeneous and similar in appearance to adjacent normal liver, while those with intratumoral hemorrhage or intracellular fat produces distinguishing imaging features.

US: Lesions with a high lipid content or hemorrhage may be hyperechoic to the normal liver; however, in the background of diffuse fatty infiltration or glycogen storage disease, adenomas may be hypoechoic compared with the remainder of the liver

Color Doppler: may demonstrate central vessels with a triphasic pattern or a continuous flat venous waveform with no central arterial flow. Peripheral peritumoral arterial and venous waveforms may be seen.

CT: it is typically sharply defined, with a pseudocapsule seen in 25%-30% non enhanced scan shows hypo dense lesion, areas of hyper density may be noted with recent hemorrhage in about 15%-43%. They may be heterogeneous with areas of lipid or fat seen at CT in 7%-10% and calcification in 5%-15%.

Enhancement pattern depend on the tumor size: the small lesions < 4 cm, are typically homogeneously enhancing, while the large lesions heterogeneously enhance because of necrosis, fat, hemorrhage, and calcification. It usually demonstrates preferentially hepatic arterial enhancement and isoattenuating in the portal venous and delayed phases (Fig.15).

MRI adenomas are hyperintense to the normal liver on T1wi and T2-weighted images. T1 hyperintensity may be attributed to intracellular glycogen or lipid demonstrates signal dropout on opposed-phase or fat-suppressed images in 36%-77% of cases. Yet, this finding is not specific for adenoma, as 40% of hepatocellular carcinomas also histologically contain fat. T1 hyperintensity can also be due to hemorrhage in 52%-93% of cases. T2 hyperintense areas may pseudocapsule may be seen that is hypointense on T1wi, is variable on T2wi, and may enhance.

1. Focal Nodular Hyperplasia
FNH is a benign epithelial liver tumor, most often seen in adult women but uncommonly occurs in young children and adolescents. This lesion demonstrates a complex architecture, with well-differentiated hepatocytes forming nodules subdivided by fibrous septa, which coalesce to form a characteristic central vascular stellate scar.

FNH represents 2% of all primary hepatic tumors in children from birth to age 20 years, it is typically diagnosed between the ages of 2 and 5 years. A marked female predominance of the lesion is reported.

Clinically : FNH commonly is asymptomatic and incidenta. Symptoms of a mass lesion are described in 20% of cases. Abdominal pain is another common symptom. Tumor rupture and hemorrhage are rare. AFP levels are not elevated.

### Imaging Features

The radiological appearance of FNH reflects its pathologic features. It is composed predominantly of hepatocytes, so it appears similar to normal liver. The presence of the central scar may aid identification of the mass on nonenhanced scan. The vascular supply from the hepatic artery causes early contrast enhancement relative to the adjacent parenchyma. The scar often demonstrates delayed enhancement as contrast material diffuses into the myxomatous stroma.

**US:** FNH appears as a homogeneous, well-circumscribed mass that may be iso-, hypo-, or hyperechoic. The central scar appears hyperechoic relative to the remainder of the mass. Calcification is rare. Color and power Doppler evaluation of the mass reveals increased blood flow in the central scar extending to the periphery in a spoke-wheel pattern. Spectral analysis of intratumoral flow reveals arterial waveforms, a finding that distinguishes FNH from hepatocellular adenoma, in which intratumoral flow is venous.

**CT:** non enhanced scan, FNH is typically well circumscribed and iso- to slightly hypovattenuating to uninvolved liver, and the scar is hypovattenuating

Arterial phase: FNH typically demonstrates early, uniform enhancement more than the adjacent liver in the arterial Portal and delayed scan: FNH becomes isoattenuating to the liver. Enlarged feeding arteries may be apparent on arterial phase images.

The stellate scar is typically hypovattenuating on early contrast-enhanced images and demonstrates enhancement on delayed images (Fig.16&17).

Atypical features may be observed including lack of the central scar, rapid washout of contrast material in the portal venous phase, lack of enhancement of the central scar on delayed images, early draining veins, and partial peripheral rim-like enhancement on delayed images.
**MRI** :- T1 :- FNH typically appears homogeneous and iso- to slightly hypointense to the liver, the scar is usually hypointense to uninvolved liver.

T2WI :- iso- to slightly hyperintense , scar is hyperintense too owing to edema within the myxomatous tissue of the scar.

The dynamic imaging showing the same enhancement pattern as the dynamic CT scan.

Atypical MRI features include lack of a scar, a scar that is hypointense on T2wi , a T1-hypointense enhancing pseudocapsule due to compression of surrounding parenchyma with mild fibrosis, and a strongly hyperintense lesion on T2wi or diffusely hyperintense lesion on T1wi.

99mTc sulfur colloid imaging:- FNH typically exhibits normal uptake in 60%-75% of lesions owing to the presence of Kupffer cells. The abundance of Kupffer cells is variable in FNH, and the remainder of the lesions show either increased or, less commonly, decreased radiotracer accumulation relative to normal liver parenchyma. Normal or increased uptake of colloid by the mass distinguishes FNH from hepatic adenoma and malignant tumors .

1. **5. Hepatobiliary LCH**

Hepatobiliary involvement occurs mainly in multisystem LCH. It is seen in 50%-60% of children with multisystem disease but in only 14.4%-18% of those affected by LCH generally. Langerhans cells directly infiltrate the periportal regions of the liver, showing a marked affinity for the bile ducts. Radiologic findings of liver involvement reflect the underlying histopathologic process, which comprises four phases of progression, from an initial proliferative phase to granulomatous, xanthomatous, and, finally, fibrous phases.

Hepatic involvement can include periportal infiltration (Fig.16 ), tumor-like or cystic lesions( Fig.17 ) , or overall hepatomegaly (Fig.18 ) .

Periportal inflammation with edema, which appears as bandlike or nodular areas of relative hypoechogenicity at US, hypattenuation at CT (Fig ), and moderate to high signal intensity at T2-weighted MR imaging. Associated periportal contrast enhancement may be suggestive of portal triaditis.

Primary or secondary sclerosing cholangitis may produce extra- and intrahepatic biliary irregularities with segmental narrowing and focal areas of slight dilatation, resulting in a beaded appearance of the bile ducts at conventional cholangiography, ERCP, or MRCP.
Differential diagnosis include:- Other diffuse infiltrating liver diseases, such as lymphoma, leukemia, or hepatitis. The main differential diagnosis of sclerosing cholangitis in children includes underlying inflammatory bowel disease. Cholangiopathies caused by infectious agents such as cryptosporidium or HIV, ischemia, or mechanical causes, with the development of secondary sclerosing cholangitis, are less common.

1. **6. Inflammatory myofibroblastic tumor**

Inflammatory myofibroblastic tumor (IMT) is a rare neoplasm of mesenchymal origin, primarily in soft tissues seen in many different anatomical locations. Inflammatory pseudotumor was first observed in the lung and described by Brunn in 1939 and was so named by Umiker et al in 1954 because of its propensity to clinically and radiologically mimic a malignant process.

Hepatic involvement by inflammatory pseudotumors was first described in 1953 by Pack and Baker (24). The majority of hepatic inflammatory pseudotumors occur in children and young adults. Most cases have been solitary solid tumors, mainly arising from the right hepatic lobe. In a few cases, inflammatory pseudotumor has involved the porta hepatis or bile ducts, which results in obstructive jaundice.

Patients generally present with a mass or nonspecific symptoms, including vague abdominal pain or gastrointestinal complaints for intraabdominal lesions, and cough, chest pain, or, less often, haemoptysis for pulmonary tumours.

The hepatic IMFT mass appears heterogeneous echogenicity on sonography.

Low attenuation and low attenuation with a peripheral enhancing rim on contrast enhanced CT images. (Fig. 19).

The other form of hepatic IMFT appears as ill-defined, heterogeneously enhancing infiltrative lesion at the porta hepatis, with adjacent intrahepatic bile duct dilatation on CT images.

**Images for this section:**
1 year old male with hepatoblastoma

Large right hepatic lobe calcified mass

Fig. 1
Multiple variable sized heterogeneously enhancing masses in both hepatic lobes showing necrotic areas.
Left hepatic lobe heterogeneously enhancing mass showing small necrotic areas and small calcification

Fig. 3
Fig. 4

16 year male with embryonal sarcoma

Large hepatic mass with heterogeneous enhancement and small calcification, appear as a solid mass in US and cystic one in CT.
13 year old female with embryonal rhabdomyosarcoma

Multiple necrotic hepatic masses with marginal enhancement

Fig. 5
Multiple hepatic focal lesions with periportal infiltration
Fig. 7

17 years female with NHL

Large left hepatic lobe mass with intra hepatic biliary radical dilatation
5 year male with lymphoma

Multiple hepatic focal lesions with marked periportal infiltration

Fig. 8
1 year old male with Atypical teratoid rhabdoid tumor

Large right hepatic lobe mass with multiple necrotic areas and heterogeneous enhancement

Fig. 9
3 year male with metastatic Wilms' tumor

Large left renal mass with hepatic metastasis

Fig. 10
2 months male with Neuroblastoma (stage 4S)

Bilateral adrenal masses with multiple hepatic and splenic metastases

Fig. 11
3 months male with hemangioendothelioma

Multiple hepatic masses with ring enhancement and central necrosis

Fig. 12
Fig. 13

1 year female with mesenchymal hamartoma

Large solid hepatic mass with cystic areas
1 year male with mesenchymal hamartoma

Large cystic hepatic mass with enhancing solid component

Fig. 14
Fig. 15

4 year old male with adenoma

Right hepatic lobe mass with intense enhancement
Fig. 16

7 years old female with LCH

Dilated intra-hepatic biliary tract with peribiliary sheets
4 years old male with LCH

cystic dilation of the biliary tree with intrabiliary solid nodules.

Fig. 17
Fig. 18

3 years old female with LCH

Hepatosplenomegaly, diffuse hypo dense liver
A well defined oval shaped hepatic focal lesion is seen at segment 6. It is seen less enhanced than the normal hepatic parenchyma. It shows hypo intense signal on T2wi MRI and hypoechogenicity on US. Pathological DX : IMFT
Fig. 20

1 year male with Focal nodular hyperplasia

Left hepatic lobe focal lesion with intense enhancement
Fig. 21
2 year female, biliary cystadenoma

Large cystic hepatic mass with enhancing fibrous septae

Fig. 22
3 year male with calcified stromal tumor

Right hepatic mass with marginal enhancement and central necrosis

Fig. 23
ALL with FUNGAL infection

Multiple hepatic and splenic hypodense focal lesions

Fig. 24
Conclusion

Focal hepatic lesions in child encompass wide range of differential diagnosis; the awareness of the different radiological appearance of these lesions is of utmost importance for proper diagnosis and management thus better life of such children.

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References