Hepatic Cysts and Abscesses

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81.1 Hepatic Cysts

81.1.1 Simple Non-Neoplastic Cyst

These cysts are also known as congenital hepatic cysts. They are uncommon in children and are often identified as incidental findings during imaging procedures. Cysts are unilocular with fine septations and contain serous fluid. Hepatic cysts do not communicate with bile ducts. Large cysts produce atrophy of the adjacent hepatic tissue but the vessels and ducts are spared. Protruding vessels give a convoluted appearance to the cyst lining. Small cysts do not compress adjacent hepatic tissue. Intracystic bleeding can change the morphological appearance of the cysts. The lining of the cyst wall is composed of an inner layer of cuboidal or columnar epithelium and an outer wall, which contains a thin layer of connective tissue and compressed liver tissue. Most congenital hepatic cysts are asymptomatic. However, large cysts have been reported to cause respiratory distress and hydroureteronephrosis. Haemorrhage into the cyst or rupture into the peritoneal cavity causes abdominal pain. These cysts may be detected on routine prenatal screening. Hepatobiliary iminodiacetic acid (HIDA) scans and magnetic resonance cholangiopancreatography (MRCP) can be performed to distinguish them from choledochal cysts and cystic biliary atresia. Asymptomatic cysts do not require treatment. In the case of large cysts, surgical excision is warranted. Surgical techniques used in adults, such as cyst deroofing and packing of the cyst cavity with omentum, have also been performed in children. At laparotomy, the cyst fluid may be examined for biliary communication. Laparoscopic deroofing of the cyst is also a safe and effective method of treatment. Cyst resolution has been reported following percutaneous injection of ethanol into the cyst.

81.1.2 Fibrocystic Disease of the Liver

These include a group of congenital disorders such as polycycstic liver disease, congenital hepatic fibrosis, choledochal cysts, biliary hamartomas and Caroli's disease. Abnormal development of the ductal plate leads to duct dilation and cyst formation.

81.1.2.1 Polycystic Disease

Liver cysts can occur in patients with autosomal dominant polycystic kidney disease. In children, the kidney is more severely affected than the liver and although liver cysts have been reported, complications are rare. Liver involvement is characterized by numerous cysts spread through the

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liver parenchyma. They rarely present before puberty and treatment is reserved for symptomatic patients and is generally supportive of liver and kidney function. Ultrasonography shows numerous thin walled hypoechoic cysts. Liver transplantation is advised in advanced disease.

Fifty percent of children with autosomal recessive polycystic disease of the kidney will have liver involvement in the form of hepatic fibrosis. However, large cysts in the liver are rare. Most children have symptoms of portal hypertension because of hepatic fibrosis.

81.1.2.2 Congenital Hepatic Fibrosis

This condition occurs in various syndromes and presents with the clinical features of portal hypertension. It is caused by the maldevelopment of small size interlobular bile ducts. The intrahepatic bile ducts are dilated and tortuous and portal fibrosis is present. Dilated bile ducts mimic liver cysts. The outcome is good if the portal hypertension is well controlled.

81.1.2.3 Biliary Hamartoma

These are characterized by ductal plate malformations of the small intralobular bile ducts. They are rare and patients have multiple focal similar size lesions spread throughout the liver. CT scan shows hypoattenuated lesions, which are non-contrast enhancing. They can coexist with simple hepatic cysts and polycystic liver and kidney disease.

81.1.2.4 Caroli's Disease

Caroli's disease is very rare and presents in infants and young adults. This condition is characterized by multiple cystic dilations of the intrahepatic bile ducts. It is caused by ductal plate malformations of the large intrahepatic bile ducts. Segmental bile duct dilation is a variant of this disease. It affects only one part of the liver and is rare in children. Children present with recurrent episodes of cholangitis and cholelithiasis. Ultrasonography demonstrates multiple anechoic intrahepatic bile lakes. Treatment includes broad spectrum antibiotics for cholangitis. Surgical management to relieve obstruction involves removal of stones and hepatico jejunostomy or external drainage. Localized disease can be managed by lobectomy, but liver transplantation is warranted in severe cases**.** Caroli's disease is also included as a part of the classification of choledochal cysts, which have been dealt with elsewhere.

81.1.3 Cystic Neoplasms of the Liver

Intra hepatic hemangioma, previously referred to as hemangioendothelioma, present as hypoechoic lesions on ultrasound or CT scans and may be mistaken for abscesses. They are usually self-limiting and present before 6 months of age. Focal, diffuse and multifocal lesions may be seen. Multiple lesions can present with cardiac failure and hypothyroidism. Most children are successfully treated with low doses of propranolol. Large lesions may require selective embolisation or even liver transplantation. There may be small multifocal hypoechoic lesions scattered throughout the liver, which are diagnostics of multifocal hemangioendotheliomas.

Mesenchymal hamartomas are the second most common tumors of the liver in children. They present as a benign large multicystic liver mass in children younger than 3 years of age. They arise from mesenchymal cell rests that become isolated from the normal portal triad and differentiate independently. Alpha fetoprotein levels are normal. CT scans show a fluid-filled mass with internal septations and no calcification. Most tumors are amenable to surgical excision. Some tumors undergo spontaneous regression. Malignant transformation to embryonal sarcoma has also been reported.

Cystic teratomas are rare and present with calcification in the margins of the cyst. Serum alpha fetoprotein is mildly elevated in some cases. Resection is the treatment of choice because of the potential for malignancy exists especially if there are immature elements.

81.1.4 Parasitic Cysts

Hydatid disease of the liver is caused by two species, namely Echinococcus granulosus and Echinococcus multilocularis. This infection is endemic in the Middle East, Australia and South Africa. It commonly occurs between 5 and 15 years of age and the liver is involved in 60% of children. The hydatid cysts in the liver are slow growing, although in children, they grow rapidly and may appear by 3 years of age.

The parasite has a life cycle linked to the intermediate host and a definitive host. The worm lives in the intestine of animals like the dog (definitive hosts), and the ova are consumed by humans and animals like sheep and cattle who are the intermediary hosts. Humans are affected by contact with contaminated environments and infected animals.

In the intermediate host, the ova that are not destroyed by the gastric juice reach the duodenum, where the alkaline intestinal contents dissolve the membrane of the ovum thereby releasing the embryos. The embryos penetrate the bowel wall and reach the liver through the blood stream where they are filtered by the small capillaries of the liver. The embryos develop into cysts in the liver. These cysts have a pericyst layer composed of connective tissue, epithelial cells, giant cells and eosinophils. This layer is densely adherent to the liver tissue. Within the peri-cyst layer is the endocyst layer composed of an outer thick layer, which allows permeation of nutrition into the cyst and an inner germinal layer, which secretes hydatid fluid and produces scolices and daughter cysts. The hydatid fluid is very antigenic and spillage of this fluid can result in development of cysts in the peritoneal cavity.

Small cysts remain uncomplicated, while large cysts cause pressure on the adjacent liver tissue and this leads to right upper quadrant pain and mass, obstructive jaundice and cholangitis. Large cysts also cause pain and may rupture into the peritoneal cavity or into the pleural space. Sometimes there can be secondary bacterial infection in the cyst.

Asymptomatic cysts may be incidental findings during ultrasound examinations. Especially in children from endemic areas, hydatid cysts must be considered in the differential diagnosis of hypoechoic lesions of the liver.

Ultrasound scans are accurate in the diagnosis of these cysts. CT scans show calcification of the cyst wall and the presence of daughter cysts. The reliability of the Casoni's skin test is less than the indirect hemagglutination tests and the enzyme linked immunosorbent assays (ELISA) using urine or saliva.

Mebendazole and Albendazole are the drugs of choice in the treatment of hydatid disease and must be continued for 1–2 months to prevent recurrence. Surgery is restricted to children with large cysts (<5 cm in diameter) or complicated cysts that have ruptured. During laparotomy, the large cysts are aspirated and carefully opened to avoid spillage. The endocyst wall is carefully removed by blunt dissection and the residual cavity is closed with mass sutures. Cyst perforation must be treated urgently and high dose steroids in the postoperative period reduce the effects of anaphylaxis. Recently, sonography-guided percutaneous aspiration of the cyst with injection of hypertonic saline or absolute alcohol has been reported to be successful.

81.2 Hepatic Abscesses

Hepatic abscesses were initially reported in children with perforated appendicitis, but the incidence has decreased with the advent of antibiotics. However, intraabdominal infections in immunocompromised children still continue to be the source of hepatic abcesses. Bacterial infections are transmitted to the liver via the hepatic artery, bile ducts or portal veins. Most pyogenic abscesses in the western world are seen in children with immunodeficiencies like chronic granulomatous disease and those who are immunosupressed from chemotherapy and transplants. However, in Asia and Africa, pyogenic abscesses are still seen in neonates with omphalitis. In about a fifth of the children, a source for the hepatic abcess cannot be identified. Almost half of all liver abscesses occur in children younger than 5 years of age and a quarter occur during the first year of life.

Pyogenic abscesses may arise in children who have recurrent cholangitis secondary to cholelithiasis and choledochal cysts, which causes inflammation and obstruction of the bile ducts. Following a portoenterostomy for biliary atresia, intrahepatic biliomas may develop into pyogenic abscesses in the presence of secondary infection. Hematomas of the liver sustained from blunt abdominal trauma may develop into pyogenic abscesses. In generalized sepsis, bacterial seeding of the liver leads to pyogenic abscesses. Such abscesses have been reported in patients with pneumonia, osteomyelitis and endocarditis. Pyogenic abscesses develop in benign hepatic cysts following attempts at percutaneous drainage. Infection in these non-neoplastic cysts may also occur in the absence of intervention. Infection leads to an increase in size of the cysts with thickening and inflammation of the cyst wall. The commonest pathogens found in hepatic abscesses are Staphylococcus aureus, Escherichia coli, Hemophilus influenza and Pseudomonas.

Clinical features include fever, abdominal pain, jaundice, shoulder pain and dyspnoea. The symptoms may be of long duration, especially in chronic abscesses. These children also have weight loss and anemia. Diagnosis may be delayed if there is a low index of suspicion. The liver is enlarged and tender and a pleural effusion may be present. 50% of patients have a positive blood culture. Fungal cultures are positive in liver abcesses occurring in children with leukemia. Ultrasound scans are the mainstay in the diagnosis of liver abscesses. CT scans may identify abscesses missed on ultrasound scan. A chest X-ray may show elevation of the right hemi diaphragm and a right-sided pleural effusion. 70% of liver abscesses are multiple. Anemia, leucocytosis and a raised ESR are found in the majority of cases. Broad spectrum antibiotics and image guided percutaneous drainage is the treatment of choice, especially in multiple abscesses. Intravenous antibiotic therapy must be continued for 4–6 weeks, followed by oral antibiotic therapy for an additional 4–8 weeks. Drainage of the abscess, as well as percutaneous catheter placement, can be facilitated by laparoscopy. Open drainage is advocated when there is associated intestinal pathology that needs intervention or when the diagnosis is in doubt.

Neonatal liver abscesses occur in premature rupture of membrane. Rarely, they may occur in the neonates with omphalitis. They usually present as miliary abscesses and hence are not amenable to percutaneous drainage. Hence, long term antibiotic therapy is the mainstay of treatment. Septicemia in children with liver abscesses is associated with a higher mortality. Large abscesses may rupture into the subphrenic space, the pleural cavity and the peritoneal cavity, in which case these children require a drainage procedure and broad spectrum antibiotics.

81.2.1 Amoebic Liver Abscesses

Entamoeba histolytica infections are transmitted from person to person due to poor sanitation in overpopulated areas. It is endemic to Africa and Asia. Liver involvement is the commonest extra intestinal manifestation of amoebiasis. The trophozoites from the colon enter the liver via the portal vein and initiate abscess formation. Liver involvement is infrequent in children. The right lobe is more commonly affected. The clinical features include fever with rigor, weight loss and right upper quadrant pain. Dyspnoea is also seen in the presence of a right sided pleural effusion. The liver is enlarged and tender. A rapid increase in distension is an indication that rupture of the cyst is imminent.

Mild anemia, leucocytosis, raised ESR and mildly elevated liver enzymes may be present. The parasite is identified in the stool in only 30% of cases. Serological tests like an indirect hemoglobin assay is highly sensitive and specific, although PCR has become available and this may prove to be more accurate in the diagnosis. Other investigations include a chest X-ray, which may show an elevated diaphragm on the right side, pleural effusion and right lower lobe infiltration. An ultrasound scan accurately identifies the location and size of these abscesses and image-aided percutaneous aspiration to identify amoeba on smear will confirm the diagnosis. CT scan offers no advantage over ultrasound.

Most cases of amoebic liver abscesses respond to Metronidazole. With intravenous Metronidazole in doses of 35–50 mg/kg, clinical remission is seen within 72 h in a majority of cases. Radiological remission takes longer. Closed aspiration is recommended if there is no response to treatment after 4 days and in abcesses larger than 5 cms or in those located in the left lobe. Percutaneous aspiration is also advocated in rapidly enlarging abscesses or when the diagnosis is in doubt and a pus culture is required. Intraperitoneal rupture causes peritonitis and shock. In such cases, urgent surgical exploration is necessary. Rarely the abscess ruptures into the pleural or pericardial cavity.

Rupture into the pleural cavity is marked by severe dyspnea and tachypnea with clinical signs of pleural effusion. Pleural collections can be drained by a tube thoracostomy. Rupture into the pericardial space is a serious complication leading to cardiac tamponade. The pericardial effusion requires urgent decompression. Percutaneous drainage of the liver abscesses will also drain the pericardial effusion since there is a communication between both cavities. Peritoneal rupture requires open surgical drainage and lavage.

Fortunately, the mortality of the disease and the incidence of complications are on the decline due to early diagnosis and prompt treatment.

81.3 Conclusion

Several diseases phenotypically present with hepatic cysts. Hence, it is important to have an algorithmic approach to investigate and manage these cysts. Hepatic infections, which present as abscesses in the liver, also have features which distinguish among the various disease causing pathogens. Appropriate identification of the pathogen is important to institute effective treatment.

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