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Original Research Paper

Radio-Diagnosis

RADIOLOGICAL FEATURES OF HEPATIC HAEMANGIOENDOTHELIOMA IN A PAEDIATRIC PATIENT: A RARE CASE REPORT

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ABSTRACT Infantile hepatic hemangioendothelioma (IHH) is a rare benign vascular tumor that occurs in the liver of infants and young children. It is usually asymptomatic but can present with symptoms such as hepatomegaly, congestive heart failure, and consumptive coagulopathy. IHH is often diagnosed incidentally on imaging studies performed for unrelated reasons. The diagnosis can be confirmed with a biopsy or by imaging findings such as diffuse heterogeneous liver lesions with delayed enhancement. Treatment options include observation, pharmacotherapy and surgical resection. In most cases, the tumor regresses spontaneously with time. However, in rare cases, it can cause significant morbidity and mortality. Therefore, early diagnosis and careful monitoring are essential in the management of IHH.

KEYWORDS : hepatic hemangioendothelioma, vascular tumor, hepatomegaly, liver lesions.

INTRODUCTION

Infantile hepatic hemangioendothelioma (IHH) is a rare benign vascular tumor that can occur in infants and young children. Although it is usually asymptomatic, it can cause significant morbidity and mortality in some cases. The diagnosis of IHH is challenging due to its nonspecific clinical presentation and radiological findings. Radiological imaging, such as ultrasound and computed tomography (CT), plays a crucial role in the diagnosis and management of IHH. In this case report, we present a case of IHH in a 13 days old infant diagnosed incidentally on abdominal ultrasound.

The patient subsequently underwent a plain and contrast-enhanced CT scan to further evaluate the extent and characteristics of the lesion. We describe the ultrasound and CT findings and the importance of early detection and careful monitoring to prevent complications and ensure optimal patient outcomes.

Case Report

A 13 days old female child presented with abdominal distension since birth associated with poor feeding, after uneventful gestation and normal delivery. Physical examination shows palpable mass in upper abdomen. Baby was drowsy, pallid and jaundiced. Laboratory findings showed anemia with low serum iron. Coagulation studies, liver function tests and alpha fetoprotein within normal range.

Further ultrasonography and plain with contrast CT abdomen were performed. On ultrasound examination shows hepatomegaly with multiple well defined mixed echogenic lesions replacing entire liver parenchyma (fig.1).

On unenhanced CT, there was hepatomegaly with multiple diffuse hypodense (HU 25-28) lesions involving both lobes of liver without internal calcification (Fig 2). On contrast CT, Early contrast material drainage by hepatic veins and persistent peripheral enhancement during the portal venous phase were present.

There was incomplete peripheral nodular enhancement in arterial phase (fig. 3) with further lesions shows progressive peripheral enhancement and centripetal fill-in in Porto-venous phase (Fig. 4). It shows further fill in and appears hyperattenuating on delayed phase. (Fig. 5). The abdominal aorta just proximal to the celiac origin was wider than the infraceliac aorta.

Images:

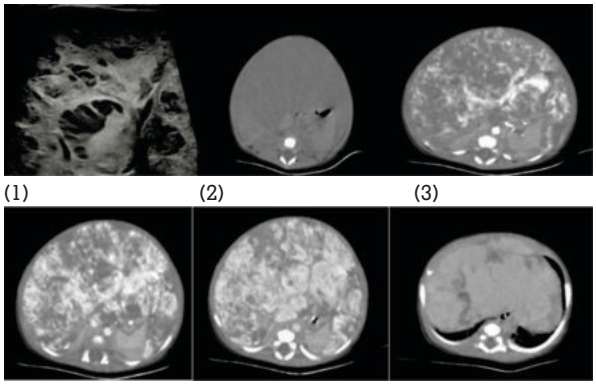

Fig. 1, Ultrasound - multiple well defined mixed echogenic lesions replacing entire liver parenchyma. **Fig 2.** Plain CT scan of axial image, Hepatomegaly with multiple diffuse iso to hypodense (HU 25-28) lesions involving both lobes of liver without internal calcification. **Fig 3**. Axial CT arterial phase- Lesions shows incomplete peripheral nodular enhancement. **Fig.4** Axial CT portal phase- lesions shows persistent peripheral enhancement with centripetal fill-in. **Fig. 5** Axial CT venous phase- lesions shows persistent peripheral enhancement with centripetal fill-in **Fig.6** Axial CT day phase- lesions shows complete central filling and appears hyperdense compare to liver parenchyma.

DISCUSSION

Infantile hepatic hemangioma is a proliferative endothelial cell neoplasm that involves the liver. The lesion is composed predominately of endothelial cells and has characteristic phases of rapid growth caused by cellular proliferation and spontaneous involution (which can be accelerated by the use of angiogenesis inhibitors). It is frequently referred to as hepatic hemangioendothelioma type 1 or type 2. [1] it may affect the liver, lungs, mediastinum, and multiple other sites. However, the most commonly involved organ is the liver.[2] The tumor has a 2:1 female predilection. Approximately 85% of affected patients present by 6 months of age [3] infantile hemangioendothelioma is the third most common hepatic tumor in children (12% of all childhood hepatic tumors), the most common benign vascular tumor of the liver in infancy, and the most common symptomatic liver tumor during the first 6 months of life [4] Most tumors continue to grow during the 1st

year of life and then spontaneously regress, probably due to thrombosis and scar formation. [5]

The clinical manifestation of infantile hemangio endothelioma is variable. The tumor may be asymptomatic and discovered incidentally. More often, however, the tumor is large and manifests as hepatomegaly, abdominal distention or a palpable upper abdominal mass. There may be extensive arteriovenous shunting within the lesion, resulting in decreased peripheral vascular resistance. Thus, increased blood volume and cardiac output are required to maintain vascular bed perfusion, which may lead to high cardiac output and congestive heart failure in up to 50%–60% of patients [6]

The imaging work-up of a child with hepatomegaly, an abdominal mass, or abdominal distention usually starts with abdominal radiography and US. Radiography may show hepatomegaly and a nonspecific mass effect in the upper abdomen with displacement of intestinal structures, as well as occasional calcifications within the projection of the liver or mass[7] At US, infantile hemangioendothelioma appears as a complex, mostly solid hepatic lesion with variable hypo- and hyperechoic echotexture. In cases of significant arteriovenous shunting, dilated hepatic vasculature with prominent blood flow at Doppler US is typical. If large vascular spaces are present, anechoic regions with detectable flow may be seen [5] At unenhanced CT, infantile hemangioendothelioma usually manifests as a well-defined mass that is hypoattenuating relative to the normal liver parenchyma[8] At contrast-enhanced CT, the enhancement pattern may resemble that of an adult giant hemangioma (4,8), with "nodular" peripheral puddling of contrast material in the early phase, subsequent peripheral pooling, and central enhancement with variable delay [3] In larger tumors, central enhancement is often lacking due to fibrosis, hemorrhage, or necrosis [9] Histologically, IHH is classified into two types. Type I IHH shows the proliferation of small, capillary-like vascular spaces lined by bland or plump endothelial cells. The vascular channels are separated by connective tissue interspersed with small bile ducts.

The mitotic figures are either rare or absent. Malignant spindle cell components are not present. Type II IHH shows disorganized branching vascular channels lined by endothelial cells, which reveal pleomorphism, nuclear hyperchromatism, and frequent mitoses [10] he endothelial cells in both tumors show positivity with CD31, CD34, and factor VIII-related antigens [11]

The size of the tumor and severity of symptoms determine the treatment modalities. Interventions are helpful in symptomatic cases which fail concerning conservative treatments. Pharmacotherapy with steroid or vincristine has been reported as the first-line treatment for IHH [12] In addition, beta-blocker, propranolol, can be used as the first-line treatment in patient with the diffuse type of IHH [12 Besides, there has been reported that the tumor shows rapid regression when adding of interferon (IFN) was made [13] surgical resections should be considered in patients with life-threatening symptoms or that the mass cannot be discerned from other malignant tumors. A natural involution of the mass can be stimulated by applying steroids and interferon [14]

CONCLUSIONS

Infantile hepatic hemangioendothelioma is one of the causes of liver mass in neonates. Physical examination, radiographic findings, and pathological findings are essential for the definite diagnosis and differentiation from other hepatic masses. Kasabach-Merritt syndrome and congestive heart failure are probable complications and should be investigated. It is important to note that surgery plays an important role in the case presenting with complications, failure of supportive treatment, or refusal of medical therapy. Prenatal diagnosis is also essential in early diagnosis, with prompt treatment after birth leading to an excellent outcome.

REFERENCES:

[1] American Journal of Roentgenology. 1975;123: 791-801. 10.2214/ajr.123.4.791
[2] Virarkar M, Saleh M, Diab R, Taggart M, Bhargava P, Bhosale P. Hepatic Hemangioendothelioma: An update. *World J Gastrointest Oncol* 2020; 12(3): 248-266 [PMID: 32206176]
[3] Roos JE, Pfiffner R, Stallmach T, Stuckmann G, Marincek B, Willi U. Infantile hemangioendothelioma. Radiographics. 2003 Nov-Dec;23(6):1649-55. doi: 10.1148/rg.236035024. PMID: 14615570.
[4] Roos, J. E., Pfiffner, R., Stallmach, T., Stuckmann, G., Marincek, B., & Willi, U. V. (2003). Infantile Hemangioendothelioma. *Radiographics*, *23*(6), 1649–1655. https://doi.org/10.1148/rg.236035024
[5] Ingram JD, Yerushalmi B, Connell J, Karrer FM, Tyson RW, Sokol RJ. Hepatoblastoma in a neonate: a hypervascular presentation mimicking hemangioendothelioma. Pediatr Radiol. 2000 Nov;30(11):794-7. doi: 10.1007/s002470000320. PMID: 11100498.
[6] Swischuk LE. Abnormalities of the liver. In: Swischuk LE, ed. Imaging of the newborn, infant and young child. 4th ed. Baltimore, Md: Williams & Wilkins, 1997; 501–521.
[7] Buonomo C, Taylor GA, Share JC, Kirks DR. Abnormalities of the hepatobiliary system. In: Kirks DR, Griscom NT, eds. Practical pediatric imaging: diagnostic radiology of infants and children. 3rd ed. Philadelphia, Pa: Lippincott-Raven, 1998; 954–979.
[8] Horton KM, Bluemke DA, Hruban RH, Soyer P, Fishman EK. CT and MR imaging of benign hepatic and biliary tumors. RadioGraphics 1999; 19:431–451.
[9] Keslar PJ, Buck JL, Selby DM. From the archives of the AFIP. Infantile hemangioendothelioma of the liver revisited. RadioGraphics 1993; 13:657–670.
[10] Zhang Z, Chen HJ, Yang WJ, et al.: Infantile hepatic hemangioendothelioma: a clinicopathologic study in a Chinese population. World J Gastroenterol. 2010, 16:4549-57. 10.3748/wjg.v16.i36.4549
[11] Kim TJ, Lee YS, Song YS, Park CK, Shim SI, Kang CS, Lee KY: Infantile hemangioendothelioma with elevated serum alpha fetoprotein: report of 2 cases with immunohistochemical analysis. Hum Pathol. 2010, 41:763-7. 10.1016/j.humpath.2009.05.019
[12] Avagyan S, Klein M, Kerkar N, et al.: Propranolol as a first-line treatment for diffuse infantile hepatic hemangioendothelioma. J Pediatr Gastroenterol Nutr. 2013, 56:17 20. 10.1097/MPG.0b013e31824e50b7
[13] Moon SB, Kwon HJ, Park KW, Yun WJ, Jung SE: Clinical experience with infantile hepatic hemangioendothelioma. World J Surg. 2009, 33:597-602. 10.1007/s00268-008-9882-4
[14] Imai H, Hidaka N, Murakami T, Kido S, Yumoto Y, Fukushima K, Kato K: In utero sonographic findings of giant hepatic hemangioma and associated perinatal complications: a report of two cases. J Med Ultrasound. 2015,23:46-51. 10.1016/j.jmu.2014.10.005