Infantile Hemangioendothelioma of Liver

Bora Sreedhar, Vijayasree Mandava, Venugopal Madubhushi, Chaganti Padmavathi Devi, Ramana Kumari Pasam
From the Department of Pathology, Guntur Medical College, Guntur, Andhra Pradesh, India.

Abstract:

Benign vascular tumors of liver are uncommon tumors and divided into infantile hemangioendothelioma and cavernous hemangioma. We report the case of a 25-day-old female child who presented with an abdominal distension. CT abdomen showed hypoattenuating mass in left lobe of liver. The mass was excised and pathologic diagnosis of infantile hemangioendothelioma of liver was made. Hemangioendothelioma of liver has been reported as most common benign symptomatic vascular tumor of the liver in infancy.

Key words: Hemangioendothelioma, Liver, Hemangioma, Child, Humans.

Introduction

Infantile hemangioendothelioma is the third most common hepatic tumor in children, seen in 12% of all childhood hepatic tumors. It is 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. Approximately 85% of affected patients are symptomatic by 6 months of age and in about 45-50% of cases these patients have accompanying cutaneous hemangiomas [1]. Two types of infantile hemangioendotheliomas have been identified on the basis of the tumor size and vascularity [2]. Solitary lesions were more common than multiple lesions [1]. This present case was a female child who presented with solitary lesion in infancy.

Case Report

A 25 days female child was admitted in the pediatric ward with abdominal distension, respiratory distress and regurgitation of milk. Our patient had no history of birth asphyxia, jaundice and fever. A definite fullness of abdomen was seen in epigastric and left hypochondric region. Palpation revealed a nonpulsatile mass in epigastric and left hypochondric region. Antenatal and perinatal history of mother was uneventful. Ultrasound of abdomen showed a solid heterogenous lesion in the left lobe of liver. Abdominal computed tomography confirmed a well defined hypoattenuating mass in left lobe of liver. Routine investigations including liver function tests were within normal limits.

Corresponding Author: Dr. Bora Sreedhar
Email: sreedhar930@gmail.com
Received: December 21, 2014 | **Accepted:** December 24, 2014 | **Published Online:** December 25, 2014

Conflict of interest: None declared | **Source of funding:** Nil | **DOI:** http://dx.doi.org/10.17659/01.2014.0130

Our patient was then scheduled for surgery. A laparotomy was performed, mass was freed from its adhesions, partial resection of left lobe of liver was done and specimen sent for histopathological examination. We received single grey brown, well circumscribed nodular mass of size 9.5x9x7 cm. Cut section was capsulated and variegated [Fig.1]. Microscopic appearance revealed tumor composed of vascular spaces of varying sizes separated by fibrous stroma. Vascular spaces are lined by single layer of cells that are cytologically bland [Fig. 2]. The case was diagnosed as infantile hemangioendothelioma of liver. The postoperative course was uneventful and patient was discharged 10 days after surgery.

Discussion

Infantile hemangioendotheliomas are usually benign, but malignant sarcomas have been reported to arise in existing hemangioendotheliomas [2]. The clinical presentation of infantile hemangioendothelioma is variable. The tumor may be asymptomatic and discovered incidentally or with hepatomegaly or a palpable upper abdominal mass. In our case the patient presented with abdominal mass. Hematologic abnormalities like leukocytosis, aneamia and thrombocytosis may be seen [3]. In the present case the hematological investigations were within normal limits. On imaging, infantile hemangioendothelioma appears as a complex, mostly solid hepatic lesion with variable hypo- and hyperechoic echotexture [4]. Radiological findings are correlating with the present case.

Grossly the tumor shows variegated appearance, small cysts, areas of necrosis, grey-white and gelatinous appearance [5]. In the present case tumor was well circumscribed and grey-white. Microscopically infantile hemangioendothelioma is composed of a connecting network of vascular channels lined by endothelial cells. Areas of varying degrees of hemorrhages, necrosis, calcification,

Fig.1(a & b): Gross photograph of the lesion showing single grey brown, well circumscribed nodular mass of size 9.5x9x7 cm. Cut-section shows variegated appearance.

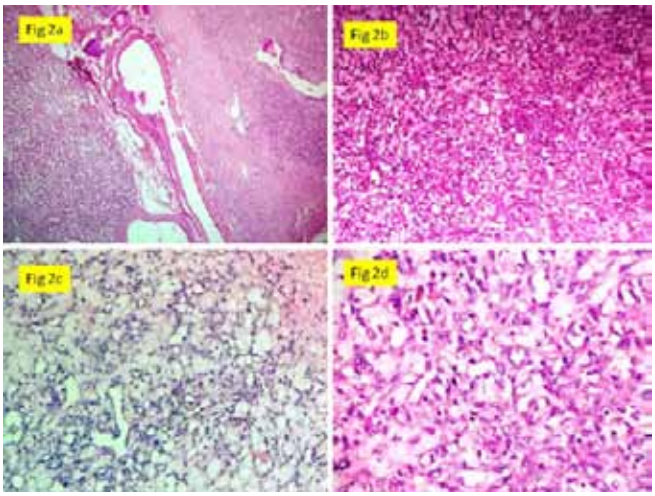

Fig.2(a): H&E 5x10, shows lesion with adjacent liver parenchyma (arrow). **(b):** H & E, 10x10, showing tumor composed of vascular spaces of varying sizes separated by fibrous stroma. **(c) & (d):** H & E, 40x10, showing vascular spaces lined by cytologically bland looking endothelial cells.

thrombosis, or fibrosis are seen in large tumors [6] .

Two types of infantile hemangioendotheliomas have been identified on the basis of the tumor size and vascularity. Type I lesions are often calcified and consist of multiple small, vascular channels with an immature endothelial cell lining and a fibrous stromal

separation containing bile ductules between the channels. Type II lesions have a more disorganized appearing endothelial cell lining and no stromal bile ductules [3]. Microscopic appearance of our case correlated with type I lesion.

Most important differential diagnosis to infantile hemangioendothelioma in this age group is hepatoblastoma, which is a malignant embryonic tumor, seen in infants and children under 5 years of age [2,7]. Patients with infantile hemangioendothelioma usually have an excellent prognosis, and spontaneous regression is known to occur after the 1st year of life. Nevertheless, children may die of associated complications such as severe heart failure [3]. Treatment is determined on the basis of the tumor size and the severity of symptoms like congestive heart failure, arteriovenous shunting or coagulopathy. Intervention is necessary only if the lesion is symptomatic and cannot be managed conservatively while the expected involution occurs. Surgical resection is indicated if life-threatening symptoms are present or if the mass cannot be distinguished from a malignant tumor radiologically [2].

Conclusion

We have reported the case of an infant with infantile hemangioendothelioma presenting with abdominal distension. The awareness of the diagnosis along with a timely diagnosis, will improve overall prognosis of ailment.

References

1. Selby DM, Stocker JT, Waclawiw MA, Hitchcock CL, Ishak KG. Infantile hemangioendothelioma of the liver. Hepatology. 1994;20(1):39-45.
2. Zenge JP, Fenton L, Lovell MA, Grover TR. Case report: infantile hemangioendothelioma. Curr Opin Pediatr. 2002;14:99-102.
3. Zhanz Zhang, Hui-Jiao Chen, Wen-Juan Yang, Hong Bu, Bing Wei, Xiao-Yu Long, *et al*. Infantile hepatic hemangioendothelioma: A clinicopathological study in a Chinese population. World J of Gastroenterol. 2010;16(36):4549-4557.
4. Ingram JD, Yerushalmi B, Connell J, Karrer FM, Tyson RW, Sokol RJ. Hepatoblastoma in a neonate: a hypervascular presentation mimicking hemangioendothelioma. Pediatr Radiol. 2000; 30:794-797.
5. Suh YL, Cho KJ, Chi JG, Park KW. Infantile hemangioendothelioma of liver - a case report. J Korean Med Sci. 1987;2(3):195-200.
6. Dehner, LP, Ishak KG. Vascular tumors of the liver in infants and children. Arch Pathul. 1971;92:101-111.
7. Keslar PJ, Buck JL, Selby DM. Infantile hemangioendothelioma of the liver revisited. RadioGraphics 1993; 13:657-667.