ACG CASE REPORTS JOURNAL

IMAGE | LIVER

Familial Presentation of Giant Liver Hemangiomas

Jalaj Rathi, MS[1], Gajendra Anuragi, MS[1], Sugaprakash Sankareswaran, MCh[1], Sugumar Chidambaranathan, MCh[1], Prabhakaran Raju, MCh[1], and Naganath Babu Obla Lakshmanamoorthy, MCh[1]

[1]*Institute of Surgical Gastroenterology, Madras Medical College, Chennai, India*

CASE REPORT

We describe a family in which 5 of its members of 2 successive generations presented with multiple giant liver hemangiomas (GLH). A 30-year-old woman in 2013 presented with abdominal pain, increasing abdominal distension, and respiratory discomfort. Multidetector computed tomography revealed multiple (8–10) hepatic hemangiomas (HH) involving almost the entire right hemiliver and caudate lobe with the largest measuring 33 × 19 × 15 cm (Figure 1), which was complicated by Kasabach-Merritt syndrome. In an attempt to reduce the hemangioma volume for safer resection, transcatheter arterial embolization of the right hepatic artery was performed but failed to cause sustained regression. Right hepatectomy was performed through a thoracoabdominal approach, without any complication (Figure 2). A histopathology report confirmed the diagnosis. She lost follow-up after 3 years and again presented in 2019 with another fresh giant hemangioma of 23 × 12 × 15 cm size arising from segments 2 and 3. Enucleation was performed successfully preserving the functional liver volume (Figure 3).

After 1 year, the patient's 43-year-old elder brother presented with abdominal discomfort and multidetector computed tomography showed multiple (4–5) hemangiomas with the largest measuring 21 × 19 × 13 cm in the right hemiliver and 11 × 9 × 6 cm in the left hemiliver (Figure 4). He underwent multiple radiofrequency ablation sessions in place of major hepatic resection, and significant relief in symptoms was achieved. We suspected familial inheritance and did ultrasound screening of the other family members, and surprisingly, of 5 siblings, the other 2 sisters and mother also had asymptomatic GLH. For the affected sisters and mother, we chose a "wait and watch" policy.

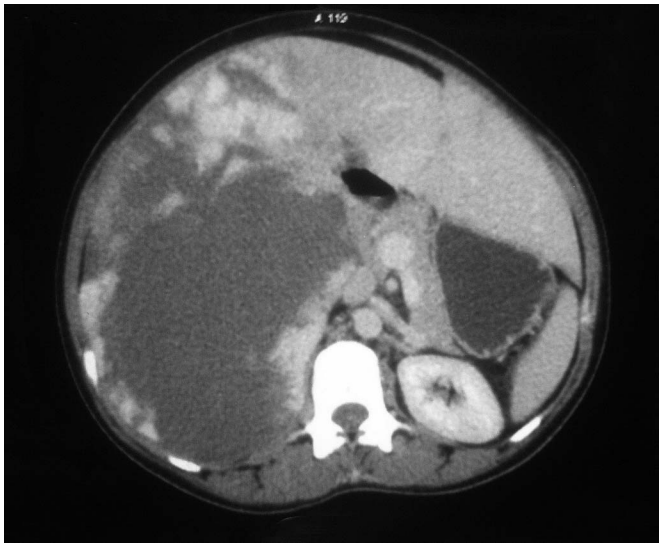

Figure 1. Multidetector computed tomography axial section showing giant hemangioma replacing the right hemiliver and caudate lobe.

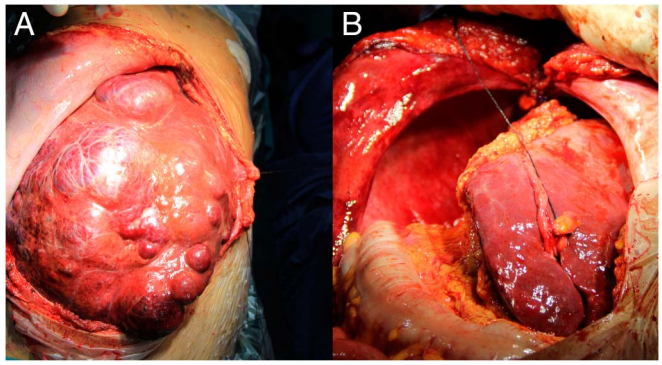

Figure 2. Intraoperative pictures revealing (A) a huge hemangioma arising from the right liver occupying almost the entire abdominal cavity and reaching up to the pelvic cavity and (B) after completion of right hepatectomy.

ACG Case Rep J 2022;9:e00765. doi:10.14309/crj.0000000000000765. Published online: July 6, 2022

Correspondence: Naganath Babu O L, Mch (naganathbabu@gmail.com).

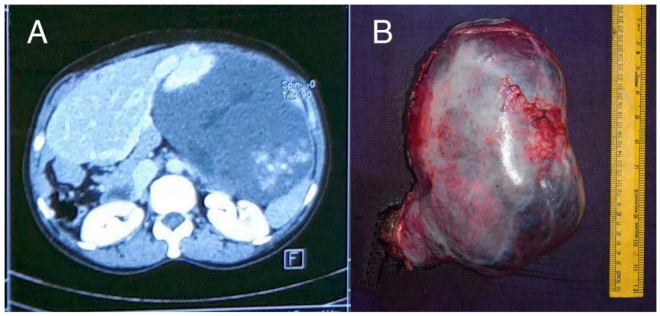

Figure 3. Newly discovered giant hemangioma after right hepatectomy: (A) multidetector computed tomography axial view displaying the tumor arising from segments 2 and 3 and (B) hemangioma specimen after enucleation.

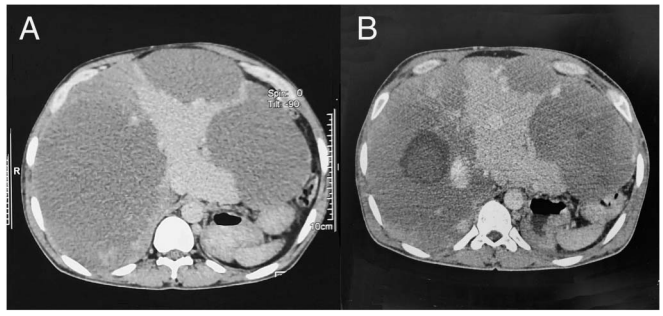

Figure 4. Multidetector computed tomography axial image of the patient's brother showing (A) multiple giant hemangiomas occupying the right and left liver and (B) after 1 session of radiofrequency ablation of right-side hemangioma.

GLH is currently defined by a diameter of 10 cm or larger.[1] The pathogenesis of GLH is still unknown. The origin of HH is likely heterogeneous and multifactorial, and there may be a genetic contribution to their development.[2] Both enucleation and liver resection are safe and effective surgical treatments for liver hemangiomas larger than 10 cm. Most GLHs are sporadic, and familial association is uncommon. We found only 3 reports of familial liver hemangioma in which an autosomal dominant inheritance with a difference in expression or penetration was proposed, but no genetic defect has been described.[3–5] Familial forms of infantile and cerebral hemangiomas are also described in the literature through the autosomal dominant inheritance. Recent research suggests that HH results from abnormal vasculogenesis and angiogenesis. We believe that identification of any genetic factor would provide insight into the pathogenesis and evolution of sporadic hemangiomas, and these rare familial forms in further research.

DISCLOSURES

Author contributions: J. Rathi wrote the manuscript. G. Anuragi provided the images. Sugaprakash S, Sugumar C, Prabhakaran R, and Naganath Babu O L revised and edited the manuscript. Naganath Babu O L is the article guarantor.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

Received July 22, 2021; Accepted November 17, 2021

REFERENCES

1. Di Carlo I, Koshy R, Al Mudares S, Ardiri A, Bertino G, Toro A. Giant cavernous liver hemangiomas: Is it the time to change the size categories? *Hepatobiliary Pancreat Dis Int.* 2016;15(1):21–9.
2. Triana P, Rodríguez-Laguna L, Giacaman A, et al. Congenital hepatic hemangiomas: Clinical, histologic, and genetic correlation. *J Pediatr Surg.* 2020;55(10):2170–6.
3. Moser C, Hany A, Spiegel R. Familial giant hemangiomas of the liver. Study of a family and review of the literature. *Praxis.* 1998;87(14):461–8.
4. Díez Redondo P, Velicia Llames R, Caro-Patón A. Hemangiomas hepáticos familiares. *Gastroenterol Hepatol.* 2004;27(5):314–6.
5. Admiraal G, Van Schie A, Van Meyel J, Vasmel W. Giant liver hemangioma in three sisters. *J Gastroenterol Hepatol.* 2004;19(3):344–5.