

Littoral Cell Angioma of the Spleen With Massive Splenomegaly Treated by Hand-Assisted Laparoscopic Splenectomy After Splenic Artery Embolization

Sung Pil Yun, MD, Jae Hun Kim, MD, Hyung Il Seo, MD, PhD, Sung Jin Park, MD

Department of Surgery, Biomedical Research Institute, Pusan National University Hospital, Busan, South Korea (all authors).

ABSTRACT

Introduction: Littoral cell angioma of the spleen is an extremely rare primary vascular tumor.

Case Description: We report a case of littoral cell angioma of the spleen with massive splenomegaly. A 37-year-old man with anemia, thrombocytopenia, leukocytosis, and splenomegaly was admitted to our hospital. A computed tomography scan of the abdomen showed multiple hypodense nodules in the spleen, and the size of the spleen was 20 × 29 cm. The preoperative computed tomography diagnosis was littoral cell angioma of the spleen. We successfully performed hand-assisted laparoscopic splenectomy after splenic artery embolization, and the final diagnosis was littoral cell angioma.

Discussion: HALS with SAE during the appropriate time period is feasible and adequate for LCA with massive splenomegaly.

Key Words: Littoral cell angioma, Hand-assisted laparoscopic splenectomy, Splenic artery embolization, Splenomegaly.

Citation Yun SP, Kim JH, Seo HI, Park SJ. Littoral cell angioma of the spleen with massive splenomegaly treated by hand-assisted laparoscopic splenectomy after splenic artery embolization. CR^{SLS} e2014.00209. DOI: 10.4293/JLS.2014.00209.

Copyright © 2014 SLS. This is an open-access article distributed under the terms of the Creative Commons Attribution-Noncommercial-ShareAlike 3.0 Unported license, which permits unrestricted noncommercial use, distribution, and reproduction in any medium, provided the original author and source are credited.

This work was supported by a clinical research grant from Pusan National University Hospital in 2014.

Address correspondence to: Jae Hun Kim, MD, Department of Surgery, Biomedical Research Institute, Pusan National University Hospital, 189 Gudeok-Ro, Seo-Gu, Busan, 602-739, South Korea. Telephone: +82-51-2407238, Fax: +82-51-2471365, E-mail: jjangmdkmdk@hanmail.net

INTRODUCTION

Littoral cell angioma (LCA) of the spleen is a rare primary vascular tumor arising from normal littoral cells lining the venous sinuses of the splenic red pulp.¹ This tumor is considered benign. About 80 cases have been reported.²⁻⁴ We describe a case of LCA of the spleen with massive splenomegaly that was successfully treated by hand-assisted laparoscopic splenectomy (HALS). Few case series of LCA treated by a laparoscopic approach have been published.⁵⁻⁷

CASE REPORT

A 37-year-old man was referred to our hospital for further evaluation of anemia, thrombocytopenia, leukocytosis, and splenomegaly. The splenomegaly and laboratory abnormalities were found incidentally during a routine checkup. The patient's medical history was unremarkable, and he was unaware of any familial-related disease. He denied having weight loss, fever, or night sweats. A phys-

ical examination showed an enlarged nontender spleen below the left costal margin. The laboratory results included microcytic anemia (hemoglobin level, 8.6 g/dL), thrombocytopenia (platelet count, $43 \times 10^3/\mu\text{L}$), and an increased white blood cell count ($12.24 \times 10^3/\mu\text{L}$). The results of blood chemistry analysis showed an elevated C-reactive protein level (4.90 mg/dL) and erythrocyte sedimentation rate (50 mm/h) and decreased levels of serum iron (17 $\mu\text{g}/\text{dL}$) and ferritin (52 ng/mL). Electrolyte levels, results of liver function tests and renal function tests, and glucose levels were normal. A computed tomography (CT) scan of the abdomen showed multiple hypodense nodules in the spleen, and the size of the spleen was 20 × 29 cm (**Figure 1**). Considering the patient's clinical symptoms and radiologic findings, our presumptive preoperative diagnosis was LCA. However, lesions with multiple hypodense nodules in the spleen have a broad differential diagnosis, including other primary vascular tumors of the spleen, other neoplastic tumors, and infection. Thus HALS was planned.

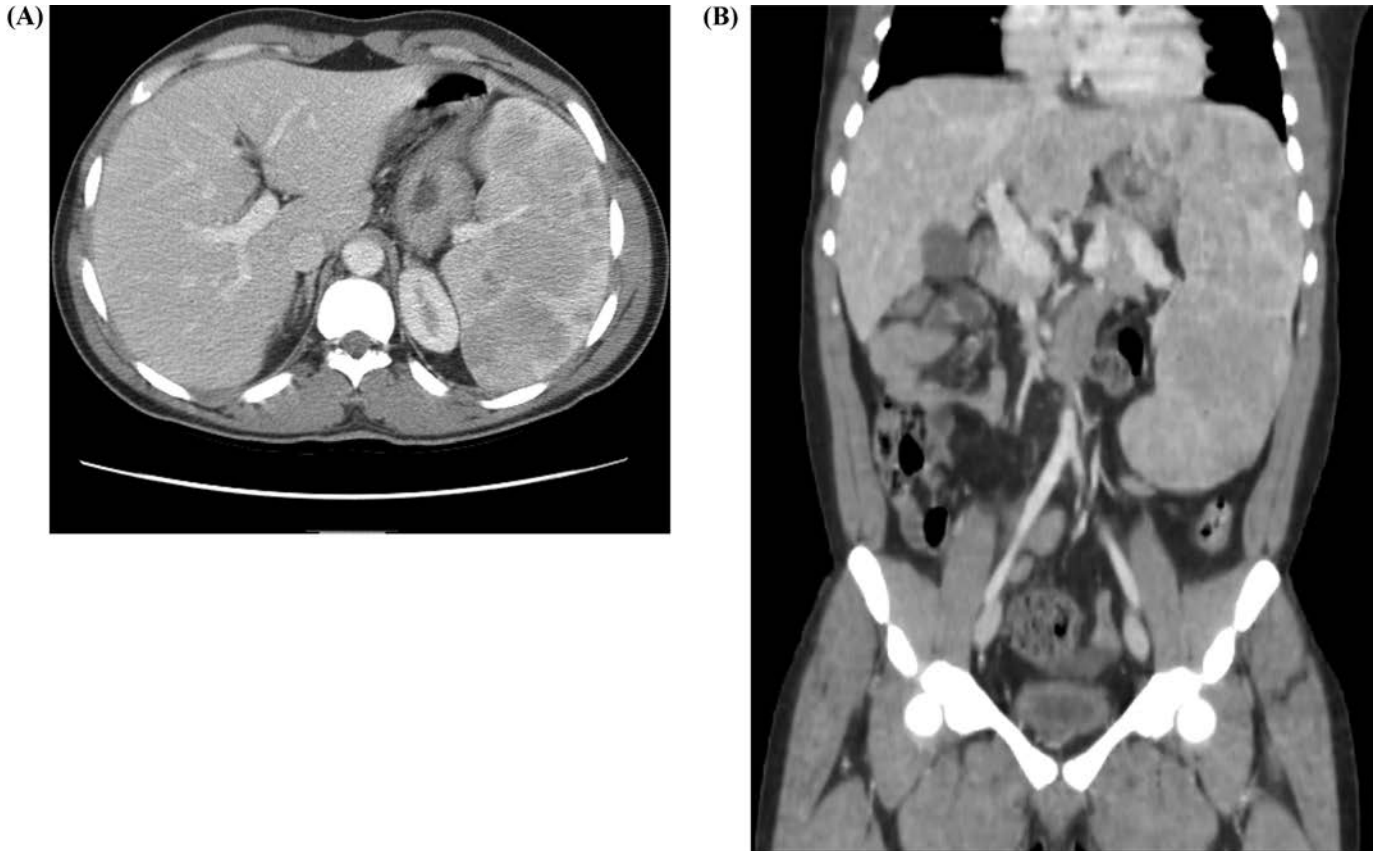


Figure 1. A computed tomography scan of the abdomen showed multiple hypodense nodules in the spleen. The size of the spleen was 20 × 29 cm.

We performed splenic artery embolization (SAE) the day before surgery to reduce intraoperative blood loss and the operative time. The SAE was performed through a microcatheter inserted into the right groin with the patient under local anesthesia. Gelfoam slurry (Ethicon, Somerville, New Jersey) was carefully injected after selection of 2 segmental arteries with a microcatheter (**Figure 2**). With the patient in the supine position, a 6-cm incision was made at the upper midline for GelPort (Applied Medical, Rancho Santa Margarita, California) to be placed. A 10-mm 30° laparoscope was placed at the superior crease of the umbilicus, and a 12-mm main operating port was placed to the right of the left midclavicular line. An additional 5-mm assisted port was set to the left of and lateral to the main port. The intra-abdominal hand provided initial dissection of the inferior pole and the splenocolic ligament, followed by dissection of the splenorenal ligament and superior pole, using electrocautery and ultrasonic devices. The splenic vascular pedicle was separated from the pancreatic tail with the intra-abdominal hand. The splenic artery was ligated with Hem-o-lok (Teleflex Medical, Re-

search Triangle Park, North Carolina), and the splenic vein was divided with an Endo GIA device (Covidien, Mansfield, Massachusetts). The intraoperative course was uneventful, but the GelPort incision was enlarged by 2 cm because an intact spleen was required for pathologic examination and the spleen was massive. The operative time was 360 minutes, and the estimated blood loss was 700 mL. The resected spleen weighed 1864.0 g and measured 28.0 × 19.0 × 8.5 cm (**Figure 3**). A spongy configuration with a few delicate gray septa and hemorrhagic features was noted in sections. Some peritumoral vascular thrombosis was associated with previous splenic arterial embolization. The lesion consisted of anastomosing vascular channels with cystic dilatation and papillary projections, and the cells lining the channels had an eosinophilic cytoplasm (**Figure 4**). The final diagnosis was LCA with hemorrhagic infarction.

The patient recovered well after surgery, but the platelet count increased gradually until postoperative day 7 because of reactive thrombocytosis. The platelet count nor-

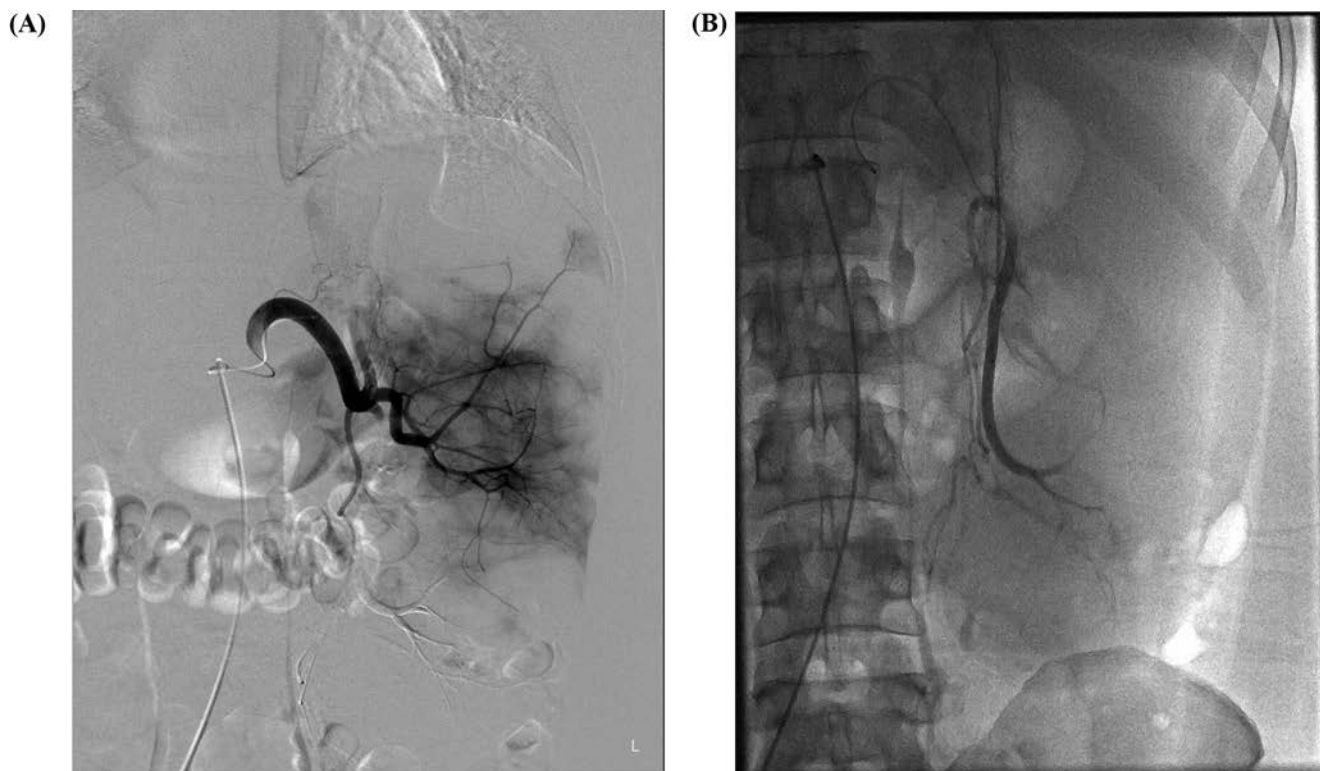


Figure 2. Preoperative splenic artery embolization was performed with a Gelfoam slurry after selection of 2 segmental arteries with a microcatheter.



Figure 3. The resected spleen weighed 1864.0 g and measured 28.0 × 19.0 × 8.5 cm in dimension.

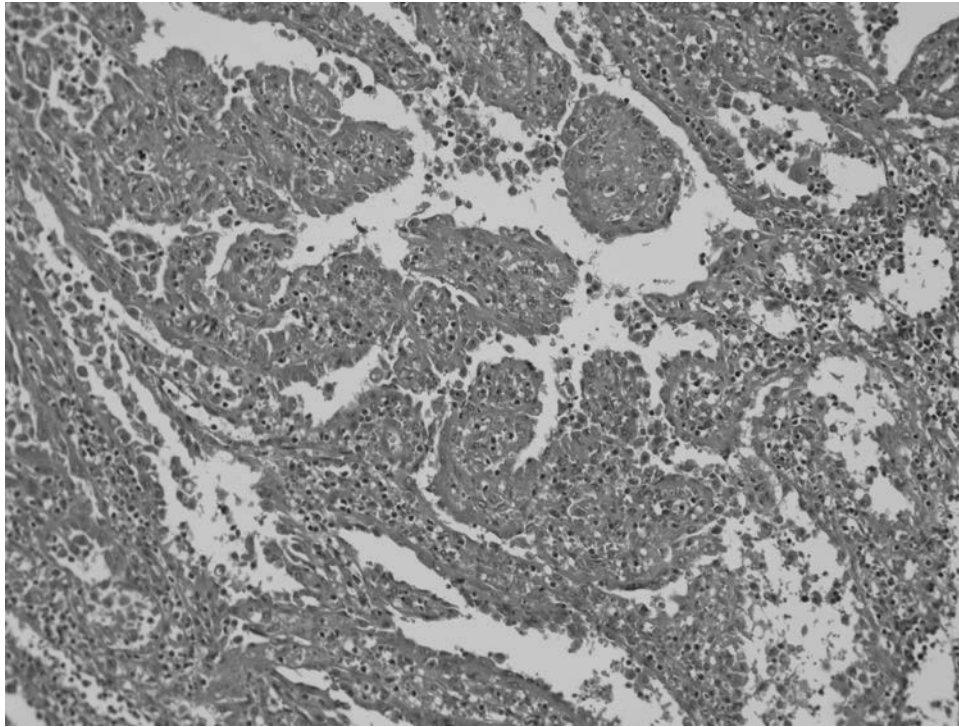


Figure 4. Anastomosing vascular channels with papillary projection and cells lining the channels showed an eosinophilic cytoplasm (H&E stain, original magnification $\times 200$).

malized after hydroxyurea therapy, and the patient was discharged on the 10th postoperative day. He was well and had no symptoms after surgery.

DISCUSSION

Primary vascular tumors of the spleen are uncommon. Most are benign, and they range from benign hemangiomas to malignant angiosarcomas.^{3,8} LCA of the spleen was reported in 1991 with characteristic features that distinguish it from other vascular splenic tumors.⁸ Gross pathologic analysis often shows a large spleen with multiple spongy red-brown nodules that are blood filled, as in our case. Littoral cells are derived from the splenic red cell pulp sinuses and have features intermediate to those of endothelial cells and macrophages. LCA is characterized by the presence of vascular channels lined by tall or flat endothelial cells with papillary projections.^{1,8-10}

Patients with LCA present with an abdominal mass due to splenomegaly, hypersplenism with ensuing anemia, or thrombocytopenia, as well as other systemic symptoms such as intermittent fever, fatigue, and weakness.^{1,9,11} Some reports have indicated that LCA may be completely asymptomatic and represent an incidental imaging find-

ing.^{3,12} Our patient had thrombocytopenia, leukocytosis, and massive splenomegaly.

The characteristics of LCA on a CT scan have been described as multiple hypoattenuating lesions with a broad differential diagnosis, including other primary vascular tumors of the spleen, other neoplastic entities, or infection.⁸ Given these CT findings, it is difficult to diagnose LCA before surgery. Levy et al⁹ reported on CT features in 8 cases of LCA that were identified as innumerable splenic masses. Our case also showed innumerable hypodense nodules in the spleen, and a preoperative diagnosis of LCA was suspected based on the patient's clinical symptoms and the CT scan.

Although LCA is considered a benign entity, close observation may be needed because of the potential for malignant change, which has been described as littoral cell angiosarcoma and littoral cell hemangioendothelioma.^{2,13} In previous reports, splenectomy was the treatment of choice for vascular splenic tumors, and symptomatic LCA was often relieved by splenectomy.^{3,14} In the present case, because the patient had thrombocytopenia and hypersplenic symptoms with massive splenomegaly, we de-

cided to perform a splenectomy, which was both diagnostic and therapeutic.

The definition of massive splenomegaly is a craniocaudal spleen length >20 cm, weight >1000 g, or both.¹⁵ The large size of the spleen decreases the working space available for dissection, making splenic mobilization more difficult.¹⁶ Studies have reported that the use of HALS has low conversion and complication rates; therefore HALS is feasible and safe for patients with massive splenomegaly.^{16–18} Wang et al¹⁹ reported that HALS for splenomegaly was associated with a shorter operative time, less blood loss, and lower conversion rate than pure laparoscopic splenectomy, suggesting that HALS was more feasible than laparoscopic splenectomy in the case of splenomegaly. In our patient HALS was inevitable because the spleen's massive size caused dense adhesion and a narrow working space for mobilization; moreover, an additional incision was required anyway for intact removal of the enlarged spleen.

Several studies have reported that preoperative embolization of the splenic artery can reduce the size of the spleen, intraoperative blood loss, and operative time.^{15,20} Poulin et al²¹ reported that SAE is used for spleens 20 to 30 cm long. In addition, Reso et al¹⁵ showed that HALS with preoperative SAE for massive splenomegaly reduces the operative time, intraoperative blood loss, and conversion rate in their comparison of previous reports on HALS without SAE. In our case we also performed preoperative SAE with a Gelfoam slurry, and the patient underwent HALS 24 hours after undergoing the procedure. Although we performed preoperative embolization of the splenic artery, we could not reduce the operative time or estimated blood loss. We assume the reason for this is the prolonged period between embolization and surgery. A previous study has indicated that the proper timing for surgery is 2 to 4 hours after embolization and a prolonged period between embolization and surgery is ineffective for reducing splenic volume in cases of splenomegaly.²² However, given the lack of studies on the proper timing of surgery after embolization, we believe that further research is required.

CONCLUSION

LCA has unique histologic and immunohistologic features, and a definite diagnosis is difficult to make preoperatively. Therefore splenectomy should be considered to relieve symptoms in patients with hypersplenism and massive splenomegaly, as well as to distinguish other malignant tumors of the spleen. HALS with SAE during the appropriate

time period is feasible and adequate for LCA with massive splenomegaly.

References:

1. Falk S, Stutte HJ, Frizzera G. Littoral cell angioma. A novel splenic vascular lesion demonstrating histiocytic differentiation. *Am J Surg Pathol*. 1991;15:1023–1033.
2. Ben-Izhak O, Bejar J, Ben-Eliezer S, Vlodavsky E. Splenic littoral cell haemangioendothelioma: a new low-grade variant of malignant littoral cell tumour. *Histopathology*. 2001;39:469–475.
3. Tee M, Vos P, Zetler P, Wiseman SM. Incidental littoral cell angioma of the spleen. *World J Surg Oncol*. 2008;6:87.
4. Hu ZQ, A YJ, Sun QM, Li W, Li L. The splenic littoral cell angioma in China: a case report and review. *World J Surg Oncol*. 2011;9:168.
5. Blansfield JA, Goldhahn RT Jr, Josloff RK. Littoral cell angioma of the spleen treated by laparoscopic splenectomy. *JLS*. 2005;9:222–224.
6. Rosen M, Brody F, Walsh RM, Tarnoff M, Malm J, Ponsky J. Outcome of laparoscopic splenectomy based on hematologic indication. *Surg Endosc*. 2002;16:272–279.
7. Yano H, Imasato M, Monden T, Okamoto S. Hand-assisted laparoscopic splenectomy for splenic vascular tumors: report of two cases. *Surg Laparosc Endosc Percutan Tech*. 2003;13:286–289.
8. Abbott RM, Levy AD, Aguilera NS, Gorospe L, Thompson WM. From the archives of the AFIP: primary vascular neoplasms of the spleen: radiologic-pathologic correlation. *Radiographics*. 2004;24:1137–1163.
9. Levy AD, Abbott RM, Abbondanzo SL. Littoral cell angioma of the spleen: CT features with clinicopathologic comparison. *Radiology*. 2004;230:485–490.
10. Bhatt S, Huang J, Dogra V. Littoral cell angioma of the spleen. *AJR Am J Roentgenol*. 2007;188:1365–1366.
11. Ziske C, Meybehm M, Sauerbruch T, Schmidt-Wolf IG. Littoral cell angioma as a rare cause of splenomegaly. *Ann Hematol*. 2001;80:45–48.
12. Harmon RL, Cerruto CA, Scheckner A. Littoral cell angioma: a case report and review. *Curr Surg*. 2006;63:345–350.
13. Rosso R, Paulli M. Littoral cell angiosarcoma: a truly malignant tumor. *Am J Surg Pathol*. 2004;28:1255.
14. Willcox TM, Speer RW, Schlinkert RT, Sarr MG. Hemangioma of the spleen: presentation, diagnosis, and management. *J Gastrointest Surg*. 2000;4:611–613.
15. Reso A, Brar MS, Church N, Mitchell P, Dixon E, Debru E. Outcome of laparoscopic splenectomy with preoperative splenic artery embolization for massive splenomegaly. *Surg Endosc*. 2010;24:2008–2012.

16. Kaban G, Czerniach D, Cohen R, et al. Hand-assisted laparoscopic splenectomy in the setting of splenomegaly. *Surg Endosc.* 2004;18:1340–1343.
17. Targarona E, Balague C, Cerdan G, et al. Hand-assisted laparoscopic splenectomy (HALS) in cases of splenomegaly. *Surg Endosc.* 2002;16:426–430.
18. Barbaros U, Dincag A, Sumer A, et al. Prospective randomized comparison of clinical results between hand-assisted laparoscopic and open splenectomies. *Surg Endosc.* 2010;24:25–32.
19. Wang KX, Hu SY, Zhang GY, Chen B, Zhang HF. Hand-assisted laparoscopic splenectomy for splenomegaly: a comparative study with conventional laparoscopic splenectomy. *Chin Med J (Engl).* 2007;120:41–45.
20. Sharma D, Shukla VK. Laparoscopic splenectomy: 16 years since Delaitre with review of current literature. *Surg Laparosc Endosc Percutan Tech.* 2009;19:190–194.
21. Poulin EC, Mamazza J, Schlachta CM. Splenic artery embolization before laparoscopic splenectomy. An update. *Surg Endosc.* 1998;12:870–875.
22. Iwase K, Higaki J, Yoon H-E, et al. Splenic artery embolization using contour emboli before laparoscopic or laparoscopically assisted splenectomy. *Surg Laparosc Endosc Percutan Tech.* 2002;12:331–336.