Fan et al., Surgery Curr Res 2012, 2:3
DOI: 10.4172/2161-1076.1000117

Surgery: Current Research

Research Article Open Access

Laparoscopic Splenectomy in the Treatment of Hereditary Spherocytosis

Ying Fan, Shuodong Wu*, Yang Su, Jing Kong and Yu Tian

Department of the Second General Surgery, Sheng Jing Hospital of China Medical University, Shenyang City, China

Abstract

Aim: To discuss the feasibility and therapeutic effect of Laparoscopic Splenectomy (LS) in the treatment of Hereditary Spherocytosis (HS).

Methods: A total of 6 patients who underwent LS for HS from August 2003 to June 2009 at our institution were recruited: 3 male and 3 female with a median age of 16.2 (range 14–24) years. The clinical data were collected and analyzed retrospectively.

Results: All the 6 operations were successful without conversion to open surgery. The average operation time was 150 minutes, the average blood loss was 46 ml, the average fasting duration was 2.25 days and the average hospital stay was 6.5 days. The jaundice began to fade 3 days after operation averagely. The symptoms of anemia were improved and the mean level of hemoglobin increased from preoperative 83.6 (range 55-105) g/L to postoperative 124.2 (range 110-140) g/L. All the patients were free of postoperative complications.

Conclusions: LS in the treatment of HS is safe and feasible with the advantage of earlier recovery and less complication compared with open splenectomy.

Keywords: Laparoscopic splenectomy; Hereditary spherocytosis; Treatment outcome

Introduction

Hereditary spherocytosis (HS) is an autosomal dominant inheritance disorder of the red blood cell membrane that presents with hemolytic anemia. Its clinical features include anemia, jaundice, and splenomegaly, increased number of small, spherical mature erythrocytes and increased permeability of the red blood cells. Erythrocyte membrane structural defect is an important mechanism in etiology of HS, and is mainly due to lack of spectrin in the erythrocyte cell membrane, leading to insufficient formation of spectrin tetramer, with a relative increase of spectrin dimers. This results in cytoskeletal weakness and instability, giving the red cells a spherical appearance, with stiffer, less malleable cell membranes that are destroyed in the spleen [1,2]. This condition is congenital and no current therapy can achieve reduction of spherical red blood cells. The spleen is the main organ where erythrocytes are destroyed, and although splenectomy does not solve the congenital defect, it stops pathological hemolysis, prolonging the red cell life span, and is therefore, the only effective treatment available [3].

Some hematological disorders, such as secondary thrombocytopenic purpura, which is not associated with splenomegaly, can be managed effectively with laparoscopic splenectomy (LS) [4,5]. In this case, LS is safe and effective, with minimal injury, less intra-operative blood loss, fewer complications on the incisions, short hospital stay, less pain and facilitates examination for accessory spleen [6,7]. Reports of LS in the treatment of HS associated with splenomegaly are relatively rare [8,9]. This study reports 6 cases of HS successfully treated with LS in our department from August 2003 to June 2009.

Materials and Methods

Patients

6 patients composed of 3 male and 3 female, aged 14 to 24 years, had LS. 4 of the patients had a clear familial history. Laboratory examination of blood smears showed more than 20% microcytic spherocytosis in all 6 cases; reticulocytes less than 5% in 1 patient, 5%-20% in 5 patients; increased erythrocytes fragility in 5 patients and normal in 1 patient; hemoglobin electrophoresis and Coomb's test were both negative; bone marrow examination of the erythroid cell line demonstrated different degrees of active hyperplasia, mainly in the mid and late stage erythroblasts. Other cell lines were normal. Mean length of follow-up was 32.6 (range 24-95) months. Clinical and laboratory parameters collected included age, sex, pre- and postoperative laboratory values (including hemoglobin concentration; bilirubin level), splenic size, operation time, blood loss, postoperative abdominal pain, fasting duration, occurrence of postsplenectomy sepsis, mortality, average hospital stay , and presence of cholelithiasis on ultrasound.

Operative method

The patient was kept under general anesthesia in a supine position, with the left flank elevated about 30 degrees. Trocars were placed at the umbilicus (10 mm), below the xyphoid process, lower third of the line between the xyphoid process and the umbilicus (both 5 mm), and the intersection between the left mid-clavicular line and a horizontal line at the level of the umbilicus (12 mm) (Figure 1).

The procedure began by freeing the lower pole of the spleen from its loose surrounding tissue; the ultrasound scalpel was used to dissect the splenicolic, splenorenal and splenophrenic ligaments. The stomach was retracted to the right with atraumatic forceps to expose the gastrosplenic ligament which was then cut, take care to avoid injury to the short gastric vessels. The porta spleen was dissected to expose the splenic artery and vein by gradual dissection from the superior border of the pancreas. The splenic artery was first ligated (Figure 2) and the spleen given time to shrink, after which the splenic vein was also ligated (Figure 3). The proximal ends of the blood vessels of the porta spleen were ligated using twin silk ligature no.7 and double titanium clips. The

***Corresponding author:** Shuodong Wu, Sheng Jing Hospital of China Medical University, Shenyang City, China, Tel: 086-024-96615-1-31211; Fax: 086-024-96615-1-31211; E-mail: coolingpine78@163.com

Received March 28, 2012; **Accepted** April 28, 2012; **Published** April 30, 2012

Citation: Fan Y, Wu S, Su Y, Kong J, Tian Y (2012) Laparoscopic Splenectomy in the Treatment of Hereditary Spherocytosis. Surgery Curr Res 2:117. doi:10.4172/2161-1076.1000117

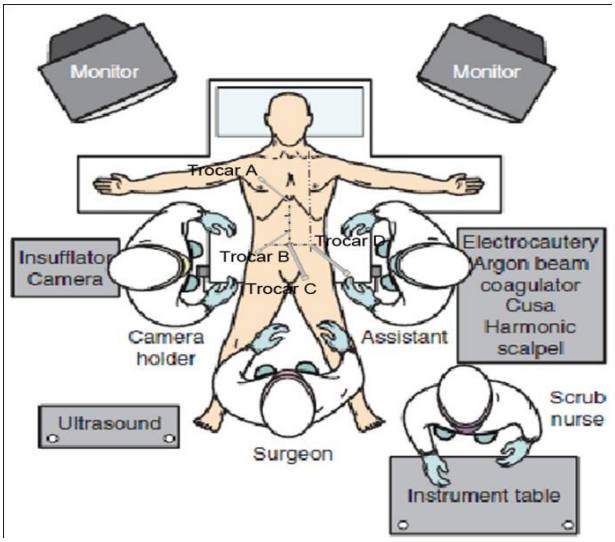

Figure 1: Team set-up and placement of the trocars. Trocar A: below the xyphoid process. B: lower third of the line between the xyphoid process and the umbilicus. C: at the umbilicus. D: the intersection between the left mid-clavicular line and a horizontal line at the level of the umbilicus.

Figure 2: Ligation and occlusion of the spleen artery.

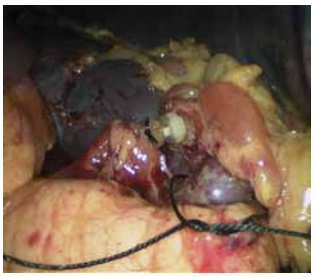

Figure 3: Ligation of the spleen vein.

distal ends were similarly ligated before the vessels were severed. The spleen was placed in an endobag and its mouth withdrawn through the left lower abdominal incision after appropriate extension. An oval clamp was used to crush the spleen in the endobag after which it was extracted piecemeal. Care should be taken to avoid puncturing the endobag resulting in transplantation of splenic tissue. The abdominal cavity was cleaned with saline and inspected for bleeding, integrity of the ligatures, presence of accessory spleen, and injury of the neighboring gastric wall and colon. An abdominal drain was placed and the incisions closed.

All the patients received antibiotics and standard thromboprophylaxis (low weight molecular heparin calcium injection 0.4 ml/day) after surgery.

Results

All the 6 patients in this series had successful LS with no conversion to open surgery. Operative time was 120 to 220 minutes, mean 150 minutes. Intra-operative blood loss was 20 to 100 ml, mean 46 ml. The mean size of the pathological spleens was 18 cm × 11 cm × 9 cm (range from 16 cm × 10 cm × 8 cm to 20 cm × 12 cm × 10 cm). Postoperative pain was assessed by visual analog scale [10]. The postoperative pain scores of the patients on day1 were all 1/10 (Table 1).No patient required ICU care after the operation, and patients were able to resume oral diet after 2 to3 days, mean 2.25 days. The abdominal drains were removed after 3 to 5 days, mean 3.75 days. Jaundice regressed after 2 to 4 days, mean 3 days. The symptoms of anemia were improved and the mean level of hemoglobin increased from preoperative 83.6 (range 55-105) g/L to postoperative 124.2 (range 110-140) g/L. Patients were ready for discharge 6 to 7 days after the operation (mean 6.5 days). The patients were followed up for an average of 32.6 months, during which no post operative infections, surgical site complications or presence of cholelithiasis on ultrasound occurred.

Discussion

The reported incidence of HS for a general population is 1 in 2000 to 1 in 5000. Although the diagnosis of HS is often made in childhood and young adult life, it may be diagnosed at any time of life including old age [11]. HS is an autosomal dominant inheritance disorder that can affect both male and female subjects. In almost all cases it is heterogeneous, with a clear familial history. A few sporadic cases related to gene mutations are also known to occur [12]. In this series, four subjects had a clear family history of HS. In one case, a parent (mother) had history of anemia and splenomegaly treated through open splenectomy.

Splenectomy is very effective in reducing haemolysis, leading to a significant prolongation of the red cell life span. The clinical manifestations and complications (anaemia and gallstones) are much reduced in severe HS and abolished in milder cases. However, concerning over the possible consequences of sepsis has led some groups to investigate whether it is necessary to remove the whole spleen to control haemolysis. Partial splenectomy is theoretically associated with a decreased risk for post-splenectomy sepsis, but it is possible that further surgery may need to be undertaken for either recurrence of haematological problems or for symptomatic cholelithiasis [13]. In this small series study, we performed LS for all the patients and no postoperative sepsis occurred.

Should a concurrent cholecystectomy be performed during LS is still in controversy. There is general consensus that individuals with symptomatic gallstones should undergo cholecystectomy, although there are no randomized trials examining this question. If stones are

	operation time(min)	blood loss(ml)	postoperative pain socres	hospital stay(d)
LS	150 ± 37	46 ± 28	1	6.5 ± 1.5
Conventional open splenectomy[a]	186 ± 45	100 ± 22	3.5 ± 0.5	9.5 ± 1.5
P value	<0.05	<0.05	<0.05	<0.05

[a]Conventional open splenectomy for HS: a retrospective analysis of the perioperative data of the last six non-complicated cases undergone conventional open splenectomy for HS dated before 1 Aug 2003

Table 1: comparison betwwen LS and open splenctomy for HS.

an incidental finding without symptoms, the value of cholecystectomy remains controversial [14]. In this study, all the patients had no gallstones before operation. Therefore, no cholecystectomy was performed. Close follow-up after the operation showed no presence of cholelithiasis on ultrasound.

It is common for HS to be associated with splenomegaly. We therefore recommend that a right side lie position is adopted for the patient. Also, the position of the patient can be adjusted as necessary during the operation: it is convenient to slant the patient 30 degrees to the right to enhance exposure and clearance of the short gastric vessels and splenic attachments, and when packing specimen into the endobag; slanting 60 degrees exposes the posterior of the diaphragm; in cases of massive splenomegaly, clearing the suspensory ligaments of the spleen first leaves it hanging on its own weight, therefore intra-operative adjustment of the patient's position enhances exposure and manipulation. Because the lower pole of the spleen is shifted inferiorly in splenomegaly, the observation port needs to be adjusted respectively to maintain a functional field of view.

The management of the blood vessels in the splenic pedicle is a key step in successful LS. In our experience, the splenic pedicle is best dealt with in the following ways: 1. the splenic artery and vein should be ligated first. In this way the splenic pedicle can be controlled to avoid bleeding. Splenic blood flow also reduces making the spleen to shrink in size, creating more room for manipulation [15]. 2. After ligating the splenic artery, the splenic pedicle should be disconnected after change in color of the spleen is observed. This is necessary as it is possible to confuse large branches for the main splenic artery thereby causing heavy bleeding when the pedicle is disconnected. 3. When using the ultrasound scalpel, Ligasure or Endo-GIA to secure the splenic pedicle, care should be taken to ensure the full circumference of the pedicle is secured to avoid leakages.

In conclusion, our experience has shown that placing the patient in the right side position makes LS safe and feasible even for HS patients with enlarged spleen. Advantages of LS include reduced risk for complications, lower risk of bleeding, shorter hospital stay and quicker recovery. Improved surgical techniques coupled with better equipment and increasing experience of surgeons in performing LS will contribute to the safety and widespread use of this operation.

References

1. Eber S, Lux SE (2004) Hereditary spherocytosis--defects in proteins that connect the membrane skeleton to the lipid bilayer. Semin Hematol 41: 118-141.
2. Ozawa K (2008) [Pathophysiology, diagnosis and treatment of anemia]. Nihon Rinsho 66: 423-428.
3. Bolton-Maggs PH, Stevens RF, Dodd NJ, Lamont G, Tittensor P, et al. (2004) Guidelines for the diagnosis and management of hereditary spherocytosis. Br J Haematol 126: 455-474.
4. Ozdemir A, Karakoc D, Hamaloglu E, Kologlu M, Ozenc A (2004) Laparoscopic splenectomy for haematological diseases. Acta Chir Belg 104: 555-558.
5. Chowbey PK, Goel A, Panse R, Sharma A, Khullar R, et al. (2005) Laparoscopic splenectomy for hematologic disorders: experience with the first fifty patients. J Laparoendosc Adv Surg Tech A 15: 28-32.
6. Balagué C, Targarona EM, Cerdán G, Novell J, Montero O, et al. (2004) Long-term outcome after laparoscopic splenectomy related to hematologic diagnosis. Surg Endosc 18: 1283-1287.
7. Sapucahy MV, Faintuch J, Bresciani CJ, Bertevello PL, Habr-Gama A, et al. (2003) Laparoscopic versus open splenectomy in the management of hematologic diseases. Rev Hosp Clin Fac Med Sao Paulo 58: 243-249.
8. Petersen CR, Bulut O, Jess P (2008) [Laparoscopic splenectomy in children with hereditary spherocytosis]. Ugeskr Laeger 170: 925-926.
9. Choi YS, Han HS, Yoon YS, Jang JY, Kim SW, et al. (2007) Laparoscopic splenectomy plus cholecystectomy for treating hereditary spherocytosis combined with cholelithiasis in siblings. Minim Invasive Ther Allied Technol 16: 317-318.
10. Wewers ME, Lowe NK (1990) A critical review of visual analogue scales in the measurement of clinical phenomena. Res Nurs Health 13: 227-236.
11. Perrotta S, Gallagher PG, Mohandas N (2008) Hereditary spherocytosis. Lancet 372: 1411-1426.
12. Delaunay J (2007) The molecular basis of hereditary red cell membrane disorders. Blood Rev 21: 1-20.
13. Morinis J, Dutta S, Blanchette V, Butchart S, Langer JC (2008) Laparoscopic partial vs total splenectomy in children with hereditary spherocytosis. J Pediatr Surg 43: 1649-1652.
14. Shao T, Yang YX (2005) Cholecystectomy and the risk of colorectal cancer. Am J Gastroenterol 100: 1813-1820.
15. Palanivelu C, Jani K, Malladi V, Shetty R, Senthilkumar R, et al. (2006) Early ligation of the splenic artery in the leaning spleen approach to laparoscopic splenectomy. J Laparoendosc Adv Surg Tech A 16: 339-344.