Laparoscopic splenectomy for hereditary spherocytosis.

A case series and review of the literature

Ann Ital Chir, 2018 89, 6: 569-571
pii: S0003469X18028701
free reading: www.annitalchir.com

Florin Zaharie, Mihai-Stefan Muresan, Ciprian Tomuleasa, Gheorghe Popa
Collaborators: Cristina Blag, Roxana Zaharie, Claudiu Zdrehus, Mihnea Dragomir, Horia Bumbea, Alexandru Irimie, Delia Dima

General Surgery Department, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj Napoca, Romania

Laparoscopic splenectomy for hereditary spherocytosis. A case series and review of the literature

Hereditary spherocytosis (HS) is a common inherited hemolytic anemia caused by a defective erythrocyte cellular membrane. Irrespective of type of surgery performed, several case reports comparing the two type of procedures, have not proven any significant difference between serum bilirubin, serum hemoglobin, red blood cells' and platelets' count, in the follow-up period at 6 and 12 months respectively, even if platelet count has maintained high for the first 6 months postoperatively, justifying an oral antiplatelet therapy for this time interval. In the present work, we present the use of LS as the treatment of choice for HS as a case series, with all the characteristics.

KEY WORDS: Hereditary Spherocytosis, Laparoscopic Splenectomy

Background

Hereditary spherocytosis is a common inherited haemolytic anemia caused by a defective erythrocyte cellular membrane. Out of all of causes leading to inherited hemolytic anemia in the pediatric population, hereditary spherocytosis (HS) is one of the most common (1:2000 births) [1]. Familial inheritance, in an autosomal recessive manner (75%) or sporadic mutations (25%) are the most frequent encountered manners of dissemination of HS. Patients in hemolytic crisis are treated with blood transfusions, whereas more severe cases such as recurrent hemolytic crisis, severe aplastic crisis, cholelithiasis or developmental delays are treated with a splenectomy.[2,3] Ever since Delaitre and Maignien described laparoscopic splenectomy [4], this procedure has been the standard-of-care in most European hematological centers, as already proven by Rogulski *et al* [5], that have compared the results of laparoscopic splenectomy (partial *vs.* total) in children with spherocytosis. They have concluded that operating and hospitalization times do not differ between partial and total splenectomy, with blood tests showing similar recovery times in both types of surgical interventions.

Case Report

Our case series representing a single-center Central European experience with 11 patients treated with total splenectomy for hereditary spherocytosis, the results were slightly better in comparison with the Polish experience. They report a median operating time of 2:50 to 4:45 whereas we report an operating time of 1:29 to 2:10. The hospitalization time is reported to be between 7 to 17 days whereas in our experience it is between 2 and 5 days. The recovery time after the surgery is much better because of the shorter operating time, in patients that have similar characteristics as the one in Rogulski *et al* experience.

Pervenuto in Redazione Aprile 2018. Accettato per la pubblicazione Giugno 2018.
Correspondence to: Mihai-Stefan Muresan, M.D., Department of Surgery, Iuliu Hatieganu University of Medicine and Pharmacy, Cluj Napoca, Romania (e-mail: mihai.stefan.muresan@gmail.com)

ABBREVIATIONS

HS – hereditary spherocytosis
LS – laparoscopic splenectomy

Irrespective of type of surgery performed (total LS or PLS), several case reports comparing the two type of procedures, have not proven any significant difference between serum bilirubin, serum hemoglobin, red blood cells' and platelets' count, in the follow-up period at 6 and 12 months, respectively [6], even if platelet count has maintained high for the first 6 months postoperatively, justifying an oral antiplatelet therapy for this time interval. Platelet levels have normalized in most of the case series reported at 12 months [2,3]. On the other hand, if PLS has been the primary treatment of choice, secondary total LS was needed in a long-term follow-up cohort, if increased reticulocyte levels have been encountered within the first two years postoperatively, signifying recurrent HS [7,8]. In the present work, we present the use of LS as the treatment of choice for HS as a case series, with all the characteristics, as well as presenting the surgical technique in a filmed video (Supplementary material - Video filming of the laparoscopic approach).

Discussion

Due to the modified character of the red blood cells (microspherocytes) mechanical destructions of the cells in small caliber vessels is frequently encountered. In the majority of the cases, these modified red blood cells are degraded in the spleen. Acute hemolytic anemia, requiring emergent blood transfusions, is rarely the clinical scenario of HS, but in such situation, associating emergency laparoscopic splenectomy (LS) has proved to be very efficient [9]. Splenectomy for hereditary spherocytosis is a viable therapeutical option, with good results reported worldwide. The outcome for the complex management of a spherocytosis patient is due to the functionality of the medical teams, that includes the surgeon, the hematologist and the pediatrician. Still, the duration and accuracy of the surgical intervention may play an important role on the therapeutic for these patients. In all other cases, of mild to severe HS associated to splenomegaly, hypersplenism, recurrent need for blood transfusions, catastrophic aplastic statuses, associated choledocholithiasis or a delay in physical development are encountered, LS has become the standard of care in the management of HE non-responsive to standard pharmacologic therapy [5].

To our best knowledge, no centralized specific guidelines for the surgical management of HE is available. The Society of American Gastrointestinal and Endoscopic Surgeons (SAGES) in its published guidelines for LS, has made specific recommendations only regarding laparoscopic surgery in pregnant women.[10] Since HS is most frequently diagnosed at a young age, concerns regarding contacting an Overwhelming Post-Splenectomy Infection (OPSI) – related to encapsulated bacterial species (*Streptococcus pneumoniae, Neisseria meningitidis and Haemophilus influenzae*) in the postoperative setting have been raised (1-5% of cited cases) [11-13]. Thus, several centers have recommended to perform a partial laparoscopic splenectomy (PLS) leaving behind an immunological active splenic remnant of either 10 cm^3 or of about 20-30% of the splenic volume, or even adapted to the patient's body mass index (BMI) [2,14,15]. If PLS is performed, the splenic remnant still holds immunological competence without major hemolytic comorbidities [16], but if its volume is still high, pulmonary hypertension or early onset of atherosclerotic have been cited as long-term complications [15]. In the majority of cases where PLS has been the chosen approach, the upper splenic pole was preserved with the upper splenic vessels left intact. Postoperative volumetric reassessment should be undertaken via abdominal ultrasound or CT scan. The intrahilar branching of the splenic vessels sometimes makes it difficult to dissect the upper pole's vasculature and in such cases, preserving only the short gastric vessels supplying the upper pole of the spleen could offer a solution, with higher rates of splenic infarctization in the postoperative setting, requiring re-operation. If ever uncertain of the status of the short gastric vessels, the surgeon should consider converting to a total LS [16].

Conclusion

Except for emergency situations, if total LS is considered, preoperative management should consist of standard triple-vaccinations against Streptococcus pneumoniae, Neisseria meningitidis and Haemophilus influenza and meningococcus, associated to anemia correction via blood transfusions, where needed. Primary and postoperative levels of total bilirubin, serum hemoglobin, red blood cells' and platelets' counts, followed by reticulocyte count, should be routinely performed. In order to optimally plan the surgical approach (total LS or PLS), an angio-CT scan is recommended to be performed, assessing the splenic vasculature, splenic size and short gastric vessels status. Further investigations should consist of at least an abdominal ultrasound with splenic volumetry, with or without a MRI assessing the biliary tract, each and every time chole/choledocholithiasis is suspected, or acute hemolytic episodes have been recurrent [17-20]. If such is the case, most of the surgeons opt for a concomitant laparoscopic cholecystectomy (LC), without significant differences concerning patient's positioning or repositioning on the operative table or to which organ

(spleen/gall bladder) should be approached first. Doppler ultrasound scans are recommended to be performed if PLS has been the opted, in order to determinate the blood supply of the remnant spleen.

Riassunto

La sferocitosi ereditaria (HS) è una anemia emolitica ereditaria in relazione alla presenza di una membrana eritrocitaria difettosa. Indipendentemente dal tipo di chirurgia adottata, numerose esperienze riportate il letteratura paragonando le due tecniche - splenectomia totale o parziale – non hanno dimostrato differenze significative tra tassi di bilirubinemia, emoglobinemia, conta eritrocitaria e piastrinica nel periodo postoperatorio a 6 e 12 mesi rispettivamente, anche se la conta delle piastrine si è mantenuta elevata nei primi 6 mesi postoperatori, giustificando un trattamento orale antipiastrinico per tutto questo periodo.
In questo studio viene presentata una serie di splenectomie laparoscopiche quale trattamento di scelta, con tutti I dati relativi.

Author contributions

All authors were involved in the clinical management of the patient. Mihai Muresan, Florin Zaharie and Ciprian Tomuleasa wrote the manuscript. Gheorghe Popa supervised the manuscript, corrected it and approved the final version.

References

1. Iolascon A, Andolfo I, Barcellini W, et al. :*Recommendations for splenectomy in hereditary hemolytic anemias*. Haematologica. 2017; 102(8):1304-313.

2. Vasilescu C, Stanciulea O, Tudor S, et al. Laparoscopic subtotal splenectomy in hereditary spherocytosis : to preserve the upper or the lower pole of the spleen? Surg Endosc. 2006; 20(5):748-52.

3. Vasilescu C, Stanciulea O, Tudor S. Laparoscopic versus robotic subtotal splenectomy in hereditary spherocytosis. Potential advantages and limits of an expensive approach. Surg Endosc. 2012; 26(10):2802-9.

4. Delaitre B, Maignien B. Splenectomie par voie coelioscopique. Observation. Presse Med. 1992; 20(44):2263.

5. Rogulski R, Adamowicz-Salach A, Matysiak M, et al. :*Laparoscopic splenectomy for hereditary spherocytosis-preliminary report*. Eur J Haematol. 2016; 96(6):637-42.

6. Vecchio R, Intagliata E, Ferla F, Marchese S, Cacciola RR, Cacciola E: *Laparoscopic splenectomy in patients with hereditary spherocytosis: Report on 12 consecutive cases*. Updates Surg. 2013; 65(4):277–81.

7. Manciu S, Matei E, Trandafir B:*Hereditary spherocytosis - diagnosis, surgical treatment and outcomes. A literature review*. Chirurgia (Bucur). 2017; 112(2):110-16..

8. Rosman CWK, Broens PMA, Trzpis M, Tamminga RYJ: *A long-term follow-up study of subtotal splenectomy in children with hereditary spherocytosis*. Pediatr Blood Cancer 2017; e26592.

9. Winslow ER, Brunt LM:*Perioperative outcomes of laparoscopic versus open splenectomy: a meta-analysis with an emphasis on complications*. Surgery 2003; 134(4):647-53-5.

10. *National Guideline Clearinghouse Guidelines for diagnosis, treatment, and use of laparoscopy for surgical problems during pregnancy [Internet]. 2007 p. 1–25. Available from: https://www.sages.org/publications/guidelines/guidelines-for-diagnosis-treatment-and-use-of-laparoscopy-for-surgical-problems-during-pregnancy/*

11. Davies JM, Lewis MPN, Wimperis J, et al. :*Review of guidelines for the prevention and treatment of infection in patients with an absent or dysfunctional spleen: prepared on behalf of the British Committee for Standards in Haematology by a working party of the Haemato-Oncology task force*. Br J Haematol 2011; 155(3):308-17.

12. Tracy ET, Rice HE: *Partial splenectomy for hereditary spherocytosis*. Pediatr Clin North Am 2008; 55(2):503-19.

13. Castagnola E, Fioredda F:Prevention of life-threatening infections due to encapsulated bacteria in children with hyposplenia or asplenia: *a brief review of current recommendations for practical purposes*. Eur J Haematol 2003; 71(5):319–26.

14. Morinis J, Dutta S, Blanchette V, et al. : *Laparoscopic partial vs total splenectomy in children with hereditary spherocytosis*. J Pediatr Surg. 2008; 43(9):1649–52.

15. Seims AD, Breckler FD, Hardacker KD, Rescorla FJ. Partial versus total splenectomy in children with hereditary spherocytosis. Surg 2013; 154(4):849-55.

16. de Buys Roessingh AS, de Lagausie P, Rohrlich P, et al. : *Follow-up of partial splenectomy in children with hereditary spherocytosis.* J Pediatr Surg 2002; 37(10):1459–163.

17. Bader-Meunier B, Gauthier F, Archambaud F, et al. : *Long-term evaluation of the beneficial effect of subtotal splenectomy for management of hereditary spherocytosis*. Blood 2001; 97(2):399–403.

18. Guaglio M, Romano F, Garancini M, et al. : *Is expertise in pediatric surgery necessary to perform laparoscopic splenectomy in children? An experience from a department of general surgery*. Updates Surg. 2012;64(2):119–23.

19. Sasaki A, Nitta H, Otuska K, et al. : *Concomitant laparoscopic splenectomy and cholecystectomy*. Surg Laparosc Endosc Percutan Tech 2010; 20(2):66-8.

20. Ruparel RK, Bogert JN, Moir CR, et al. : *Synchronous splenectomy during cholecystectomy for hereditary spherocytosis: Is it really necessary?* J Pediatr Surg 2014; 49(3):433–5.