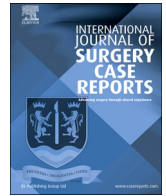




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Partial Splenectomy in the treatment of an adult with β thalassemia intermedia: A case report

João Guardado Correia^{a,*}, Nídia Moreira^b, Carlos Eduardo Costa Almeida^b,
Luís Simões Reis^b

^a Instituto Português de Oncologia de Coimbra Francisco Gentil, Av. Bissaya Barreto 98, 3000-075 Coimbra, Portugal

^b Centro Hospitalar e Universitário de Coimbra (Hospital Geral – Covões), S. Martinho de Bispo, 3041-853 Coimbra, Portugal

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ABSTRACT

INTRODUCTION: Thalassemia is a common disease which treatment is often based on splenectomy. The risks associated with total splenectomy stimulated partial splenectomy as a potentially alternative therapy.

CASE PRESENTATION: A 45 year-old female patient with long term follow-up for β thalassemia intermedia started to develop signs of hypersplenism and iron overload. A partial splenectomy was performed and was observed a marked hematologic improvement while preserving the desired splenic function.

DISCUSSION: Partial splenectomy proved to provide a persistent decrease in hemolytic rate while preserving the integrity of splenic phagocytic function, presenting itself as an effective alternative to total splenectomy. After being subjected to partial splenectomy, our patient experienced a sustained control of hemolysis and showed no signs of hypersplenism or iron overload. No splenic regrowth or infectious complications were observed. The major drawbacks of partial splenectomy are the increased risk of intra- and postoperative bleeding, splenic remnant torsion and splenic regrowth.

CONCLUSION: Partial splenectomy is an alternative to total splenectomy for the treatment of adult β Thalassemia intermedia patients avoiding the risks associated with total splenectomy.

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1. Introduction

There are many types of thalassemia, a heterogenous group of hemoglobin disorders common in the Mediterranean, Middle East, India and Southeast Asian countries [1]. β thalassemia is an inherited autosomal recessive disease with an annual incidence estimated at 1 in 100,000 throughout the world. The phenotype includes bone marrow expansion, increased function of the spleen (splenomegaly) and chronic hemolytic anemia [2,3]. More than 200 mutations were described and are responsible for the reduction or complete absence of β -globin gene expression. Without treatment, the hallmark of thalassemia syndromes is the imbalance in the α/β -globin chain ratio leading to ineffective erythropoiesis [2,3].

The clinical presentation of β thalassemia varies widely, ranging from a mild, almost asymptomatic form, to a severe and sometimes

fatal form [4]. β thalassemia intermedia is the term used to characterize individuals presenting clinical cases between light form (thalassemia minor) and severe form (thalassemia major). Red cell production in patients with β thalassemia intermedia may be adequate to sustain hemoglobin levels greater than 7 g/dL without red blood cell transfusion and render some patients asymptomatic until adulthood. Drop in hemoglobin values may occur with advancing age, during infectious periods, pregnancy, surgery, and with the development of hypersplenism, making transfusion therapy necessary [3,5].

β thalassemia intermedia treatment is based on symptoms relief, splenectomy and folic acid supplementation [2]. Transfusion therapy remains the standard treatment for the severe forms with frequency and transfusion requirement indirectly reflecting the underlying disease severity [5]. Despite being able to control the majority of the underlying physiopathologic mechanisms, transfusion therapy also contributes a great deal to secondary morbidity [5].

Common complications of β thalassemia intermedia include hypersplenism and iron overload. Hypersplenism is secondary to the development of splenomegaly, characterized by an increase in the spleen's mechanical filtering and early destruction of blood components. Iron overload is mainly due to augmented intestinal absorption of iron caused by ineffective erythropoiesis, or from

Abbreviations: Hb, hemoglobin; Hct, hematocrit; Fr, serum ferritin; MRI, magnetic resonance imaging; OPSI, overwhelming post-splenectomy infections; Plt, platelet; Ret, reticulocytes; Tbr, total bilirubin; US, ultrasonography.

* Corresponding author.

E-mail addresses: joaocorreia86@hotmail.com (J.G. Correia), nidia.moreira.22@gmail.com (N. Moreira), carloscostaalmeida@yahoo.com (C.E. Costa Almeida), lfrs.reis@gmail.com (L.S. Reis).

occasional transfusions. A direct assessment of liver iron concentration is recommended in the follow-up of these patients, either by biopsy or by a non-invasive method such as T2 MRI. [2,3,6].

Splenectomy is the main therapy in reducing hemolysis, leading to a significant rise in red cell lifespan. By eliminating the need for regular transfusions, this surgical procedure also prevents the risk of blood-born viral infections and iron overload. In addition, there is a major improvement in patients' quality of life after surgery [7].

Nevertheless, total splenectomy exposes the patient to a life-long risk of life-threatening lethal infections and to the overwhelming post-splenectomy infections (OPSI), usually caused by pneumococcal species (70–90% of OPSI) [1,7]. Although the risk of OPSI is reduced by the use of immunizations to *Streptococcus pneumoniae*, *Meningococcus* and *Haemophilus influenzae*, as well as postoperative antibiotic prophylaxis, its risk is never eliminated. Additionally, concern persists of incomplete protection by pneumococcal vaccinations, antibiotic resistance and low compliance to antibiotic prophylaxis [8]. Despite current treatment, the overall mortality rate from established causes of OPSI ranges from 50 to 70% and the interval between splenectomy and OPSI can vary from 24 days to 65 years [7].

Partial splenectomy provides a potentially effective alternative to total splenectomy, removing enough spleen to gain the desired hematologic effects while preserving the splenic function [7,9]. It is likely that contact between antigens and lymphoid cells can proceed in the remaining spleen, preserving its immune role and making partial splenectomy the best way to prevent post-splenectomy infections [7,10].

In this report, we present a case of a woman with thalassemia intermedia who was submitted to a partial splenectomy which was successful in controlling the disease without relapse on long follow-up.

This study has been reported in compliance with the SCARE criteria [11].

2. Case presentation

The case presented is a 45-year-old female patient with long standing follow-up by hematology department who was presented to the surgical department with splenectomy intent. She was diagnosed with β thalassemia intermedia in her childhood, but recently she was requiring red blood cell transfusion more often. Moreover, she was dealing with severe complications of her disease, namely heavy iron overload and hypersplenism.

In the pre-operative consultation, the advantages and disadvantages of partial splenectomy versus total splenectomy were discussed with the patient and it was decided to perform a partial splenectomy. Previously to surgery she was immunized for *Pneumococcus pneumoniae*, *Haemophilus influenza* and *Meningococcus*.

At surgery, the patient was positioned in supine position and the abdomen entered via a left subcostal incision. The spleen was partially devascularized to maintain flow from the short gastric arcades to the upper pole and the ischemic portion of the spleen was allowed to demarcate. The splenic parenchyma was transected with LigaSure Atlas™ and bleeding from the splenic bed was controlled with an Argon beam coagulator and topical hemostatic agents (Tachosil®). The goal of the surgery was to retain 20–30% of the normal splenic tissue volume, according to Diesen et al. [9] (Images 1 and 2). The postoperative course was uneventful and the patient was discharged on postoperative day eight. Histology described morphologic aspects compatible with the diagnosis of β thalassemia.

Currently with four years of follow-up the patient is doing well, maintaining stable hemoglobin levels ranging from 7.3 to 9.9 g/dL and presenting no need for red blood cell transfusions. Marked

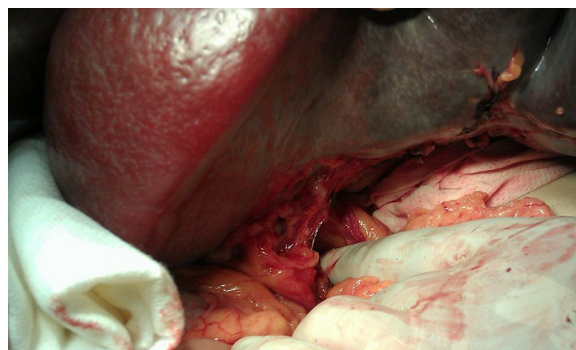


Image 1. Preservation of the short gastric arcades to the upper pole.

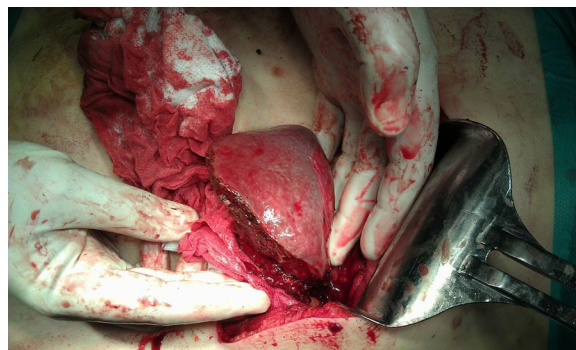


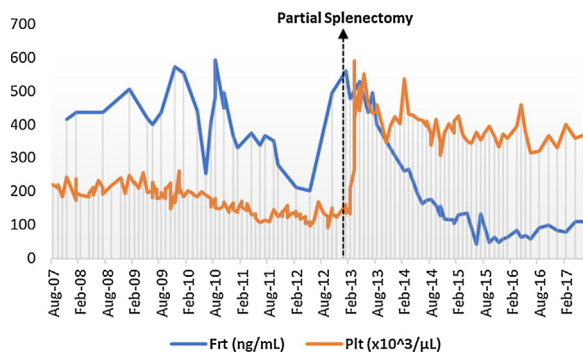
Image 2. Spleen remnant.

hematologic improvement was noticed since partial splenectomy, with sustained decrease in hemolysis parameters. Accordingly, increased levels of hemoglobin and hematocrit were observed along with reduction in the levels of reticulocyte and total bilirubin, as compared to preoperative levels (Table 2; Graphic 2). Splenic regrowth has not occurred, presenting currently a calculated splenic volume of 253.60 cc. Similarly, no signs of hypersplenism were observed, with a sustained mean platelet count increase, as compared to the preoperative levels (Table 2; Graphic 1). Iron chelation therapy was reduced to maintenance dose, without evidence of iron overload. Current hepatic iron load measured by MRI is 35 $\mu\text{mol/g}$ and there was a continuous decline in serum ferritin level (Table 1; Graphic 1). No infectious complications were observed during the follow-up period.

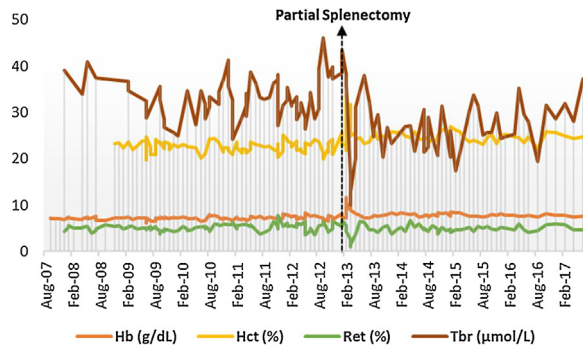
$$\text{SpleenVolume(cc)} = \text{Length(cm)} \times \text{Width(cm)} \times \text{Height(cm)} \times 0.52$$

3. Discussion

Current indications for splenectomy in β thalassemia intermedia include growth retardation, leukopenia, thrombocytopenia, increased transfusion demand and symptomatic splenomegaly. [3]. The removal of the spleen in these patients has the role of reducing the rate of hemolysis, the risk of splenic sequestration and the symptoms of splenomegaly [9]. This procedure usually allows discontinuation of transfusion needs in the majority of patients [3,7]. Total splenectomy, however, is known to be associated with severe complications, such as OPSI, thromboembolic events and pulmonary hypertension [3]. The procoagulant effect of the anionic phospholipid exposed on the surface of the damaged circulating red blood cells may contribute to the chronic hypercoagulable state responsible for other less well established but potentially severe risks of total splenectomy. It includes an increased incidence of thrombotic events and long-term risks of hypertension, vascular



Graphic 1. Evolution of serum ferritin (Frt) and platelet (Plt) throughout follow-up.



Graphic 2. Evolution of Hemoglobin (Hb), Hematocrit (Hct), Reticulocyte (Ret) and Total Bilirubin (Tbr) throughout follow-up.

thrombosis, pulmonary hypertension and cardiovascular disease [3,12].

To overcome the total splenectomy complications, a variety of procedures aiming at immunopreservation have been advocated, including partial splenectomy [4]. The partial splenectomy is based on the knowledge of the splenic vascular pattern, which divides the spleen into a variable number of almost independent vascular territories. Partial splenectomy can be performed preserving either a short gastric artery or a branch of the left gastroepiploic artery, while ligating the main pedicle of the splenic artery and vein [7].

Partial splenectomy proved to provide a persistent decrease in hemolytic rate while preserving the integrity of splenic phagocytic function, presenting itself as an effective alternative to total splenectomy [7]. Our patient experienced these benefits during the entire follow up. She had a sustained control of hemolysis demonstrated by an increase in hemoglobin and hematocrit levels along with a decrease in total bilirubin and ferritin values, with statistical significance when compared with the preoperative values (Table 2, $p < 0.05$ by independent Student t -test). Hypersplenism signs were also controlled with the surgical procedure, as confirmed by a

Table 2

Laboratory values comparison and statistical analysis by independent student t -test.

		Mean	Std. Deviation	Independent Student t -test
Hemoglobin Hb (g/dL)	Preoperative	7,21	,38	<0,05
	Postoperative	8,12	,76	
Hematocrit Hct (%)	Preoperative	22,59	1,20	<0,05
	Postoperative	25,38	2,24	
Reticulocytes Ret (%)	Preoperative	5,27	,77	<0,05
	Postoperative	4,70	1,10	
Total Bilirubin Tbr (μmol/L)	Preoperative	33,52	4,50	<0,05
	Postoperative	26,11	6,22	
Ferritin Frt (ng/mL)	Preoperative	415,82	102,39	<0,05
	Postoperative	160,19	127,46	
Platelets Plt (x10 ³ /μL)	Preoperative	167,47	39,79	<0,05
	Postoperative	387,16	80,27	

Table 1

Imaging evaluation of the spleen throughout follow-up.

Date	Splenic volume measures by US (cc)	Hepatic Iron Load measured by T2 MRI (μ mol/g)
Before Partial Splenectomy		
09/2009		260
10/2009	689.5 ($17 \times 13 \times 6$ cm)	
06/2011	632.22 ($15.7 \times 12.1 \times 64$ cm)	
05/2012	748.8 ($16 \times 15 \times 6$ cm)	240
After Partial Splenectomy		
03/2014		160
08/2014	241.8 ($8.9 \times 5.5 \times 9.5$ cm)	
08/2015		35
06/2017	252.60 ($9.2 \times 6.6 \times 8$ cm)	

decrease in postoperative platelet count and the maintenance of a reduced spleen volume, calculated at 253.60 cc, around 34% of the initial volume. Additionally, iron overload disappeared as evident by the statistically significant decrease in postoperative serum ferritin value (Table 2, $p < 0.05$ by independent Student t -test) and in the normalization of the hepatic iron load measured by T2 MRI (35 μmol/g) (Table 1). The phagocytic function was not directly tested, however the spleen remnant remained viable on Doppler examination during the entire follow-up as well as no infectious complications were observed, probably due the preserved splenic function. Given the low rate of OPSI with proper use of immunizations and antibiotic prophylaxis, any risk reduction for partial splenectomy compared to total splenectomy would require a prohibitively large clinical trial to be confirmed [8].

The major drawbacks of partial splenectomy are the increased risk of intra- and postoperative bleeding, the possibility of splenic remnant torsion and subsequent ischemia and later the risk of splenic regrowth. According to Héry et al., splenopexy is an important step of the procedure in order to avoid torsion and necrosis of the splenic remnant, especially when the remnant is very small [10]. The bleeding risk is limited by the early control of the pedicle and the use of an electrothermal bipolar tissue sealing system (LigaSure Atlas™) in the transection of the splenic parenchyma. This approach allows a clean and non-hemorrhagic transection of the spleen and reduces the risk of bleeding. Despite our patient has not experienced splenic regrowth, this complication seems to be a tendency at 4–6 years of follow-up, raising the possibility of a secondary surgical procedure [8]. However, according to Diesen et al, this regrowth is rarely associated with the recurrence of hemolytic anemia and the need of secondary splenic resection [9]. This low incidence of secondary completion splenectomy supports the potential advantages of long-term splenic salvage.

4. Conclusion

Partial splenectomy seems beneficial in β thalassemia intermedia patients as it reduces their transfusion requirements by decreasing the hemolysis, as well as obviates the risk of post splenectomy sepsis and thrombotic events by preserving the splenic phagocytic function. The main concern associated with partial splenectomy remains the splenic regrowth with the need of a secondary total splenectomy which was not observed in our patient during the long follow-up period.

Although the majority of the studies on partial splenectomy were conducted on children [4,8–10,12,13], this procedure seems advantageous in adults as well. Partial splenectomy is a technically challenging surgery, and its role compared to a total splenectomy for the treatment of adults with β thalassemia intermedia continues to be defined. However, we believe that with this procedure the desired hematologic effects can be achieved while maintaining the splenic function and avoiding post splenectomy complications.

Conflicts of interest

Nothing to declare.

Funding

Nothing to declare.

Ethical approval

Ethical approval was not needed since this paper describes the use of a well-known technique.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

J.G. Correia: study concept and design, data collection and analysis, writing the paper, review. N. Moreira: study concept and design, review. C.E. Costa Almeida: review. L. S. Reis: review.

Guarantor

L. S. Reis.

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