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Case Series

Surgery Section

Elective Splenectomy for Haemolytic Anaemia: A Case Series

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ABSTRACT

The spleen plays an important role in immune surveillance and haematopoiesis. Elective splenectomy is sometimes indicated for excessive cellular destruction (hypersplenism) or sequestration, lymphomatous or myeloid disease, or tumours. Hereditary haemolytic anaemias are disorders with a variety of causes, including red cell membrane defects, red blood cell enzyme disorders, congenital dyserythropoietic anaemias, thalassaemia syndromes, and haemoglobinopathies. Splenectomy is one of the therapeutic approaches in the management of haematological conditions. This is case series of 12 cases who underwent elective splenectomies for haemolytic anaemias in a tertiary care centre, over a period of one year. Out of the 12 patients, 10 patients were diagnosed to have Hereditary Spherocytosis (HS) and two patients were diagnosed with sickle cell anaemia. For all patients, preoperative blood transfusion was done to bring the Haemoglobin (Hb) above 9 g/dL for elective splenectomy. Among the 10 patients with HS, six patients had concomitant cholelithiasis for which cholecystectomy was done in the same sitting and three patients developed postoperative complication which was managed appropriately with no mortality and minimum morbidity. However, more detailed studies on this subset of patients are needed for the establishment of guidelines and optimal outcomes.

Keywords: Blood transfusion, Cholelithiasis, Massive splenomegaly, Spleen

INTRODUCTION

The spleen plays a significant part in immune surveillance and haematopoiesis. It also removes intracellular inclusions (pitting) and screens aged blood cellular elements from motion. Although the spleen has significant immune functions, elective splenectomy is occasionally indicated for hypersplenism or sequestration, or tumours. Hereditary haemolytic anaemias are disorders with a variety of causes, including red cell membrane defects, red blood cell enzyme disorders, congenital dyserythropoietic anaemias, thalassaemia syndromes, and haemoglobinopathies [1]. One of the most important indications for therapeutic management in haematological conditions is splenectomy [2].

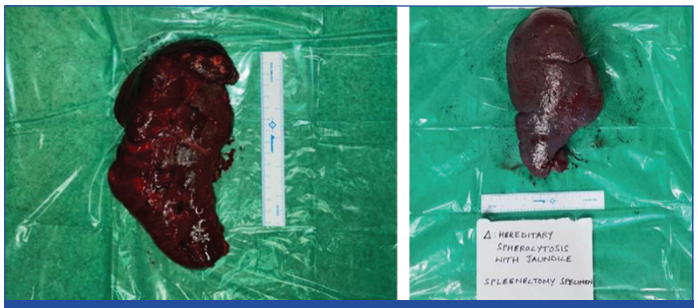
CASE SERIES

A case series of 12 patients with haemolytic anaemia was presented who were referred to the tertiary care centre for elective splenectomy over a period of one year from June 2019 to June 2020.

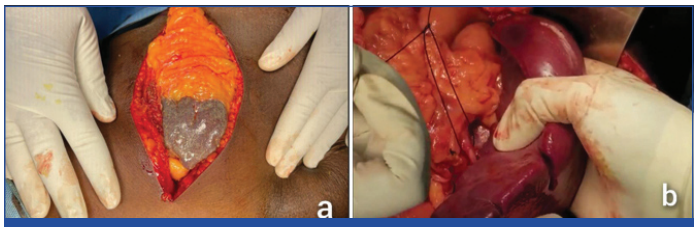
Preoperative preparation

1) All patients were properly vaccinated with Pentavalent vaccine (Diphtheria, Pertussis, Tetanus, Hepatitis B and Haemophilus influenzae type B), pneumococcal vaccine and meningococcal vaccine 72 hours before surgery.
2) Full preoperative blood work-up was done along with the cardiac status of all patients and anaesthetic fitness was obtained.
3) Preoperative Haemoglobin (Hb) of more than or equal to 9 g/dL was maintained; if found to be low, adequate blood transfusion was done before surgery.
4) Two pints of the packed cell was reserved for all patients before surgery, according to their blood groups.

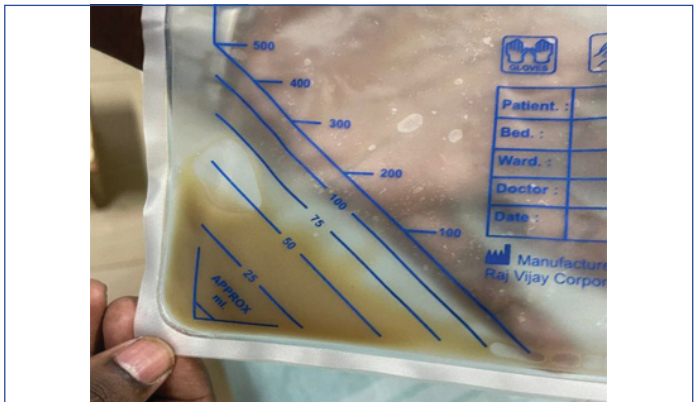
Out of 12 patients with haemolytic anaemias 10 patients were diagnosed to have HS (83%) and 2 patients with sickle cell disease (17%). For all patient elective splenectomy [Table/Fig-1-4] was done without mortality and with minimal morbidity. Out of 12 patients, six patients had concomitant cholelithiasis (50%) for which cholecystectomy was done along with splenectomy. Results have been summarised in [Table/Fig-5].



[Table/Fig-1]: Splenectomy specimen of patient with sickle cell anaemia (weight 800 gm). **[Table/Fig-2]:** Splenectomy specimen of patient with Hereditary Spherocytosis (weight 950 gm). (Images from left tor right)



[Table/Fig-3]: Intraoperative images of a patient underdoing elective splenectomy: (a) enlarged spleen seen once left subcostal incision is made; (b) ligation of splenic artery identified at splenic hilum.



[Table/Fig-4]: Drain of the patient who had pancreatic fistula with pancreatic enzyme collected in drain which was confirmed with elevated level of amylase in fluid analysis of drain collection.

Parameters		Hereditary spherocytosis (n=10)	Sickle cell anaemia (n=2)
Gender [n (%)]	Male	1	1
	Female	9	1
Mean age (in years)	Male	28	25
	Female	30	27
Elective/emergency		Elective	Elective
Open/laparoscopic		Open	Open
Mean spleen weight		860±80 gm	750±50 gm
Concomitant cholecystectomy		6 (all females)	Nil
Mean preoperative Hb (g/dL)		9.3±0.71	10±0.71
Mean operative time (min)		180±38.98	120
Mean blood loss (mL)		250±106.77	150±80
Preoperative blood transfusion		5 (1-male,4-females)	1 (male)
Preoperative complication		1 (tail of pancreas injury)*	nil
Mean hospital stays (days)		10±2.9	7
Preoperative blood transfusion		Nil	Nil
Preoperative complications		Yes (3 patients)*	Nil
Re-admission		Nil	Nil
Mortality		Nil	Nil
Mean follow-up		10 months	8 months

[Table/Fig-5]: Analysis of patients operated for elective splenectomy.
Hb: Haemoglobin; *1 patient with the tail of pancreas injury had pancreatic fistula which was treated conservatively with a retaining drain tube, till the 8th postoperative day. The remaining 2 patients had local seroma collection with wound gapping, for which secondary suturing was done in the same admission.

Postoperative management

1) Postoperative intensive care unit care was given for 3-5 days.
2) All patients were well hydrated with i.v. fluids @ 125 mL/hr.
3) Oxygen supplement was given via face mask @2-4 L for 2-3 days.
4) Deep vein thrombosis prophylaxis with inj. Fondaparinux 5 mg for 5 days.
5) Adequate postoperative i.v. antibiotics and i.v. analgesics were given.
6) Daily complete blood count monitoring for Hb and platelets was done for 3-4 days.

DISCUSSION

Hereditary haemolytic anaemias are a collection of ailments with a diversity of causes, including red cell membrane defects, enzyme disorders, congenital dyserythropoietic anaemias, and haemoglobinopathies. The definitive management has yet to be determined. Splenectomy has been recommended as a likely method to manage severely affected patients [1].

The HS is the most common form of congenital haemolytic anaemia with an incidence of approximately 1:2000 and a dominant transmission in about 70-80% of cases [3]. The risk of Overwhelming Postsplenectomy Infection (OPSI) is highest with encapsulated microorganisms though the incidence of OPSI is only 0.23% per year with a lifetime risk of 5% [4]. Strategies to reduce the development of OPSI include: (i) patient education, including advice to take urgent action in response to febrile episodes; (ii) vaccination and the advent of new protein conjugate vaccines; (iii) prophylactic antimicrobial therapy; (iv) intraoperative normothermia; (v) early catheter removal; (vi) increased vigilance [5,6]. Though these measures are thought to reduce the risk of OPSI greatly, studies reported inconsistency in implementation [7].

In this case series, a total of 12 patients were operated for elective splenectomy for a duration of one year from June 2019 to June 2020, out of which 10 patients were females and 2 patients were males. Female preponderance was seen in HS in the present series, but in studies done by other authors, it had a male preponderance [2,8-10]. The mean age of presentation was 25-28 years of age which was in concordance with the study done by other authors [Table/Fig-6]. In this series, the most common indication for elective splenectomy was HS. But in studies done by Lakhsmi BK et al., and Machado N et al., sickle cell anaemia is common, and in studies done by Rajan SP et al., and Ahmad J et al., immune thrombocytopenic purpura is the most common indication for doing elective splenectomy [2,8-10].

Studies	Present case series	Rajan SP et al., [8]	Lakshmi BK et al., [2]	Machado NO et al., [9]	Ahmad J et al., [10]
Place and year of the study	Tamil Nadu, India June 2019 to June 2020	Kerala, India October 2017 to October 2020	Andhra Pradesh, India June 2016 to June 2018	Muscat, Omen January 1993 to December 2005	Northern Ireland 1995 to 2010
No. of cases	12	42	27	150	112
Mean age of presentation (years)	Male-26.5	M-23	Female-8,	M=89	M-15
	Female-28.5	F-19	Male-19	F=61	F-17
Gender	Male-17% Female-83%	Male-54.8% Female-45.2%	Male-70% Female-30%	Male-59% Female-41%	Male:female ITP-1:2 Lymphoma 3:4 HS 3:1 AHA 2:1
Most common cause of elective splenectomy	10 cases-HS 2 cases-SCA	4 cases-HS 5 cases-Haemolytic anaemia 8 cases-ITP	23 cases-SCA 4 cases-HS	6 cases-HS 96 cases-SCA 34 cases-β-T 12 cases-ITP	13 cases-ITP 7 cases-Lymphoma 4 cases-HS 3 cases-AHA
Type of surgery	All open surgery	All open surgery	Open- 96.2% Laparoscopic-3.7%	All open surgery	All open surgery
Mean spleen weight	860 gm (HS) 750 gm (SCA)	Not available	850 gm	577 gm	Not available
Concomitant splenectomy	6 (HS)	Not available	13	13	Not available
Mean hospital stay (days)	10-HS 7 (SCA)	7	7.5	Not available	5-HS 6.5 (TTP) 7 (AHA) 10 (Lymphoma)
Complications	1 case-pancreatic tail injury 2 cases-wound gapping	Not available	4 cases-postoperative jaundice 2 cases-wound infection 1 case-thrombocytosis 1 case-pneumonia	2 cases-acute chest syndrome 1 case-fever of unknown aetiology 1 case-cavernous sinus thrombosis 1 case-intra cranial bleed 1 case-wound dehiscence	Not available

[Table/Fig-6]: Comparison of present series findings with similar previous studies [2,8-10].
HS: Hereditary spherocytosis; SCA: Sickle cell anaemia; β-T: Beta thalassaemia; ITP: Immune thrombocytopenic purpura; AHA: Autoimmune haemolytic anaemia; TTP: Thrombotic thrombocytopenic purpura

It was found that 6 patients (50%) had cholelithiasis. In studies done by Lakhsmi BK et al., 13 patients underwent concomitant cholecystectomy during elective splenectomy [2]. It has been reported that, following splenectomy, there is an increased risk of early and late venous and arterial thrombosis, including acute splenic and portal vein thrombosis, and delayed severe life-long complications [11].

It is remarkable that the rate of death from elective splenectomy in a large study is 1.6%, making it a procedure of comparable risk compared to 1-2% mortality in pancreatectomy or hepatectomy series in high-volume centres [11]. However, there was no mortality in the present study. The lesser number of patients included in the study might be the reason for this.

There is a need for the conduction of more studies on this topic with a specific focus on decision-making for surgery and perioperative protocols. In the present study, all the signs have shown improvement in the early postoperative period.

CONCLUSION(S)

Elective splenectomy is done mainly for all haemolytic anaemias with unconjugated hyperbilirubinaemia. HS is the most common cause of haemolytic anaemia.

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