Successful Innovative Minimal Invasive Splenectomy in a Child with Combined Congenital Immunodeficiency and Acquired Pancytopenia Secondary to Hypersplenism

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Abstract: Congenital combined immunodeficiency in association of acquired hypersplenism are uncommon lesions in pediatric patients and certainly poses several diagnostic and therapeutic challenges for successful and safe outcome of ideal treatment which has consequences of life long precautions. Herein, we report a case of combined immunodeficiency complicated by hypersplenism and pancytopenia in a 4-year-old female toddler who presented with hypogammaglobulinemia, low circulating B cells, and T cell lymphopenia, anemia and thrombocytopenia. The results of hematological investigations revealed pancytopenia with ongoing persistent anemia, thrombocytopenia and leukopenia. and bone marrow examination revealed adequate cellularity with normally functioning bone marrow and no circulating auto antibodies. A splenectomy using an innovative minimal invasive technique was performed for curative purposes. Any evidence of the recurrence of hypersplenism was not noted 30 months after the operation.

Surgery is the most frequently performed treatment for hypersplenism with pancytopenia and normal bone marrow and no circulating antibodies. Pre-operative and post splenectomy vaccination record and post splenectomy status cards provide additional precautions for rare but life-threatening post splenectomy complications.

Keywords: Combined immunodeficiency, Hypersplenism, Splenectomy, Status post splenectomy.

INTRODUCTION

Immunodeficiency with hypersplenism in a toddler is very challenging to manage and associated pancytopenia represent exceedingly rare lesion. Therefore, it constitutes a critical diagnostic challenge in practice, and diagnosis of these lesions is not always straightforward. Persistent pancytopenia in the presence of adequate cellularity and normally functioning bone marrow and absence of circulating antibodies should help establish the diagnosis. We strongly believe in splenic preservation in experimental, pediatric and adult population whenever feasible [1-10}. Here, we present a case of immune deficiency with hypersplenism, with a review of the literature focusing on innovative minimal invasive splenectomy and its peri-operative care. We aim to contribute to the awareness of the existence of these rare lesions and safe and successful surgical management.

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CASE PRESENTATION

A 4-year-old female patient was admitted to our children hospital with a known diagnosis of immunodeficiency with hypogammaglobulinemia, low circulating B cells, and T cell lymphopenia, anemia and thrombocytopenia. Her course was complicated and marked by multiple life-threatening respiratory infections (pneumocystic pneumonia-PCP and pseudomonas pneumonia), chronic lung disease, tracheo-bronchomalacia and severe bronchiectasis. She had undergone cord blood transplant from HLA-identical sibling, complicated by post-transplant engraftment syndrome, CMV viremia, acute kidney injury and fluid overload, and acute on chronic respiratory failure requiring intubation, hypertension, delirium, and abstinence.

At her admission, she was anemic, and a physical examination revealed hepatosplenomegaly and petechiae on the forearms. The laboratory tests showed pancytopenia with counts of red blood cells, white blood cells and platelets as shown in Figure **1A**. The patient was serologically negative for antibodies

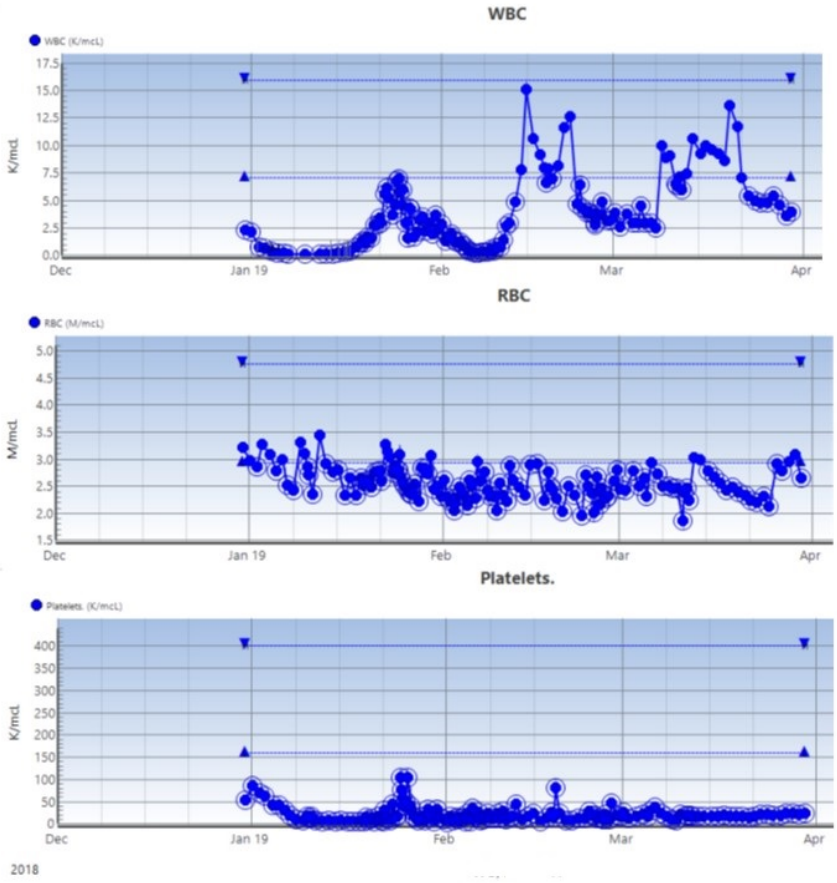

Figure 1A: Pancytopenia showing baseline white cells, red cells and platelets ongoing for last few months.

and the bone marrow showed normal cellularity and function. Abdominal ultrasonography revealed enlarged spleen and no accessory spleen. She was diagnosed to have hypersplenism and a multidisciplinary meeting indicated need for a splenectomy to improve her condition.

Pre operative vaccines were given two weeks prior to the splenectomy. Parents were given options of traditional open, innovative minimal invasive technique and laparoscopic splenectomy and they elected to go for an innovative technique developed by us over last four decades. In this technique, thorough evaluation with pre-operative abdominal ultrasound with measurements of the spleen especially lower pole, hilar region and upper pole, any detectable accessory spleen and the first rectus intersection above the umbilicus are noted. The length of the incision is equal to the lower polar length and the site of skin incision is supraumbilical centered over the first rectal intersection on the left side as marked (Figure **1B**). The rectus sheath on the right side can be opened and rectus muscle of the right side can be retracted if more room is required on the right side (especially when performing simultaneous cholecystectomy for associated gall stones) and the left side can be extended following the course of neurovascular bundle of intercostal nerves and vessels without any blood loss or injury to nerves of the abdominal wall if needed.

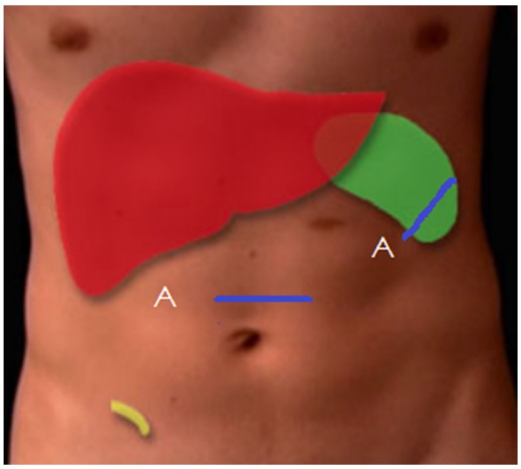

Figure 1B: Abdominal findings and measurement of lower polar splenic length and mark first left rectal intersection during ultrasonography.

The first step is to ligate the splenic artery, through a small window in the gastrocolic omental ligament, which is at the upper border of pancreas in the lesser sac followed by ligation of short gastric vessels leaving all venous drainage at the hilum of the spleen intact allowing for decompression of the congested spleen and autotransfusion of blood which is an average of two units in most cases. No blood transfusions are generally required as on an average, extra two units are pulled into the circulation and the size of the spleen comes down significantly with wrinkles observed over its surface. A search for accessory spleen and look out for any midgut malrotation with status of appendix and presence of any Meckel's diverticulum as associated anomaly are noted. Now the patient is on lighter planes of anesthesia and mainly on oxygen only at this stage and allowed some time for the spleen to decompress and gentle outside pressure over the spleen will allow quicker decompression of the congested enlarged spleen. Finally lower polar veins are ligated and divided allowing polar delivery of the lower spleen at which point hilar main splenic vein followed by upper polar venous ligation and division completes splenectomy. Final search for any accessory spleen and their removal is carried out and if any additional procedures for correcting associated midgut malrotation or pathological Meckel's diverticulum or abnormal appendix requiring any attention is sorted out at this stage and abdomen is washed with warm saline, hemostasis achieved and abdomen closed by modified single layer closure with subcuticular absorbable monofilament sutures requiring no removal and excellent cosmetic outcome.

Post operative update with vaccine record is kept with follow up vaccination in the primary care setting (Figure **2**). A splenectomy was performed using an innovative minimal invasive technique described above. The postoperative course of the patient was uneventful, and she remained well 30 months after the splenectomy.

DISCUSSION

Primary combined cellular and humoral immunodeficiency in association with hypersplenism are rare pediatric lesions requiring splenectomy. Primary immunodeficiency disorders (PIDDs) are a group of inherited conditions affecting the immune system of a child which does not function properly and cannot fight off infections, due to a problem in white blood cells, and immunoglobulin deficiency in toddlers on one hand and associated hypersplenism worsens the situation by causing pancytopenia and enhanced destruction of all three types of blood cells on the other hand posing several challenges in their management.

Spleen is an important organ for immune function and preservation of spleen is the order of the day [1-10]. Splenectomy complications include potentially lethal Overwhelming Post Splenectomy Infection (OPSI), Malaria, myocardial infarction, pulmonary embolism or pulmonary hypertension [11]. Pre-

Recommended Asplenia Vaccine Record-at least 14 days before or after surgery.

Vaccine	Date Given	Date Booster Due	Revaccination
Haemophilus B Conjugate		None	None
Pneumococcal (Prevnar-13)		None	None
Pneumococcal (Pneumovax-23)		5 years later if under age 65: _____	At age 65
Meningococcal (Menveo)		12 weeks after first dose: _____	Every 5 years
Meningococcal (Bexsero)		4-12 weeks after first dose: _____	None
Influenza		None	Every year

Figure 2: Splenectomy vaccination record card documentation.

operative vaccination is recommended 14 days prior to elective splenectomy and if spleen has been removed in emergency situation; it should be given after 14 days of splenectomy.

Splenectomy is still the most appropriate treatment option in cases of traumatic splenic rupture- grade 4 or 5 or delayed rupture following conservative management/ embolization, spontaneous rupture of pathological spleen and blood dyscrasias [11]. Pancytopenia with normal bone marrow and no circulating antibodies suggest hypersplenism; and following splenectomy red blood cells achieve a normal life span, white blood cells and platelets follows.

Partial splenectomy, auto transplant or reimplant is an option in traumatic cases while partial splenectomy can be performed in Gaucher disease and polar splenectomy is feasible in benign polar pathology [11]. Splenorrhaphy or marsupialization can be carried out in trauma cases with small lacerations or with benign cysts. Auto transplantation, partial splenectomy or reimplant of the spleen are no longer recommended in hypersplenism with pancytopenia.

Ideal age generally is at least 6 years old to reduce risk of overwhelming post splenectomy sepsis or infection (OPPS/OPSI) but it is flexible between 4 and 6 in view of pancytopenia/hypersplenism. We prefer pre-operative immunization two weeks before the planned splenectomy. Splenectomy can be done using open traditional, laparoscopic or robotic methods [12]. We developed hand assisted modified minimal invasive method using an innovative technique practiced for the last four decades combining the advantages of the open and endoscopic methods after detailed considerations of embryology, anatomy, pathophysiology, ergodynmics and combining best of open and laparoscopic methods by using the laparoscopy instruments and techniques.

Innovative splenectomy is equal or superior to traditional open and laparoscopic/robotic ones and gives best of both worlds by combining their advantages and reducing the risks and complications especially in the developing nations where resources for laparoscopic or robotic surgery and facilities for pediatric anesthesia are limited. Pre-operative mapping of the spleen size and marking of the proposed incision site gives indication of the site and size of the incision. It can be performed under laryngeal mask anesthesia thus avoiding intubation with possible post-operative respiratory complications. The ligation of arterial supply to the spleen by controlling the splenic artery and short gastric vessels and polar delivery of the spleen after its decompression are the key features allowing smaller incision. The post operative pain is significantly less with quick recovery. The scar is minimal and post operative ileus and adhesion are less common and severe. It is equally effective in the search for accessory spleen finding, control of splenic artery, short gastric and splenic hilar vessels bleeding or injury. It used for any size or weight of the spleen and can be combined with cholecystectomy. The incidence of accessory spleen is 12-32%, usually found in the hilum, along the vessels, in the greater omentum or along the splenorenal ligament. We use laparoscopic instruments such as for ligation of splenic hilum with endoscopic stapler, blunt retractor is used to elevate spleen and blunt grasper used to mimic proposed staple line for transection of hilum. Therefore, innovative approach is safe, effective, less time consuming, can be done under laryngeal mask anesthesia, have very good cosmetic and functional results, post operative pain and adhesions are less, and combining advanced technology benefits makes it speedy and accurate [14].

The incidence of portal thrombosis following pediatric splenectomy using various techniques remains the same [13]. Pre operative vaccines are required in planned splenectomy two weeks before the procedure and post operative update with vaccine record is kept with follow up vaccination in the primary care setting with an underwear and a card issued to the patient regarding splenectomy status clearly displayed and first aid measures to be instituted in case of an emergency [15].

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CONTRIBUTIONS

All authors contributed to the study conception and design. Material preparation, data collection and analysis were performed by all authors. The first draft of the manuscript was written by RP, and all authors commented on or edited previous versions of the manuscript. All authors read and approved the final manuscript.

ETHICS DECLARATIONS

Conflict of Interest

Authors Anthony, Govani Patel and Patel declare that they have no conflict of interest.

Ethical Approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent

Informed consent was obtained from all individual participants included in the study.

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