

Case Report

Laparoscopic supramassive splenectomy weighing 1710 gm in a 17-year-old boy with a history of open heart surgery, transposition of great arteries and pulmonary artery stenosis with ventricular septal defect: a case report

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ABSTRACT

When a spleen weighs more than 1,600 grams or has a craniocaudal length larger than 21 cm, it is categorized as a supramassive spleen. Laparoscopic splenectomy for normal to moderately enlarged spleens has become the gold standard in elective splenectomy surgery. Its utility in large splenomegaly has not yet completely established, but has great advantage over open surgery. Here, we present a case of 17-year-old young male with a history of congenital heart disease who underwent a Rastelli open heart surgery in 2010 and pulmonary balloon valvulotomy in 2022 came with supramassive splenomegaly, hypersplenism and pancytopenia that was treated with laparoscopic splenectomy. Vaccinations were done two weeks prior to the surgery. Preoperative embolization of the splenic artery was performed 24 hours before splenectomy to reduce complications. The excised spleen measured about 24×15 cm.

Keywords: Splenomegaly, Hypersplenism, Supramassive spleen, Laparoscopy, Transposition of great arteries

INTRODUCTION

The first laparoscopic splenectomy was performed in 1991.¹ Since then, this method has become the gold standard for elective cases involving spleens of normal or moderate size. This method has a number of advantages, including easier access to the surgical area, a quicker recovery, a reduced incidence of surgical site infection, and a lower rate of complications.²⁻⁴

Although the procedure is considered relatively safe, the outcome of treatment depends on the patient's medical history, hospital profile, surgeon's experience, and multidisciplinary approach; as seen in our case, the decision to subject a patient with congenital heart disease (CHD) to laparoscopic surgery depends on determining whether the CHD is minor, moderate, or severe.⁵

Due to the severity of the patient's condition, a multidisciplinary approach involving a pediatric cardiac surgeon, an interventional radiologist for splenic artery embolization, a cardiac anesthesiologist, and a laparoscopic surgeon was used initially.

CASE REPORT

A 17-year-old male reported with abdominal pain and lump that had been present for three months and was gradually growing in size. He had no prior history of fever, diarrhea, vomiting, or pain. He had a history of congenital heart disease (transposition of great arteries, ventricular septal defect, and pulmonary artery stenosis) for which Rastelli procedure was conducted at the age of 5 years. He had also undergone balloon pulmonary valvulotomy 3 months ago. He had no significant family history.

At the time of admission, blood tests revealed a considerable reduction in hemoglobin (7.3 g/dl), total white blood cell count (2000 cells/cumm), and platelet count (0.79 lac/cumm).

USG abdomen and MRI were prescribed as additional diagnostic procedures which revealed the presence of a 20×13 cm spleen.

In light of hypersplenism's devascularization, splenic artery embolization was performed 24 hours prior to reduce the size of the spleen and to minimize any intraoperative capsular bleed (Figure 1).



Figure 1: Size of spleen as seen on the patient's body.

After obtaining written and informed consent, a laparoscopic splenectomy was arranged for the patient. Two weeks prior to the surgery, he was vaccinated with 23-valent pneumococcal polysaccharide (PN23), the conjugated meningococcal C (MCC), and the *Hemophilus influenza* type B (Hib) vaccines.

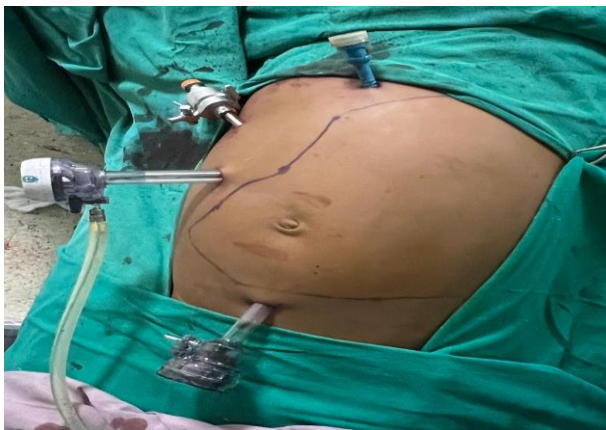


Figure 2: Ports for splenectomy.

Under general anesthesia, two 11 mm and two 5 mm ports were used (Figure 2). A low-pressure pneumoperitoneum (8 mm of Hg) was established in view of his cardiac risk instead of the standard 15 mm of Hg. First, the splenic attachments were separated, then the gastrosplenic

ligament and short gastrics ligament were ligated. Next, the splenic artery was dissected, clipped separately, and cut. Splenic vein was also dissected, ligated with no. 1 vicryl, clipped, and cut; and all remaining attachments were removed.

Hemostasis was secured and an 8cm suprapubic transverse incision was made through which the excised spleen was taken in parts (Figure 3). The excised spleen weighed about 1710 g measuring 24 cm in craniocaudal length, and 15 cm of horizontal length (Figure 4). The abdominal cavity was closed in layers, and the port sites were sutured shut. The postoperative phase was without any event.



Figure 3: Post-operative abdomen of the patient (blue arrow: old open heart surgery suture line; orange arrow: suprapubic transverse incision which was used to remove the spleen).



Figure 4: Excised spleen specimen measuring 24 cm of cranio-caudal length.

Postoperative abdominal ultrasound and arterial doppler showed no abnormal findings. During his hospitalization, the patient received two units of packed red blood cells, one pre-operatively and another intraoperatively, to treat his anemia. There was a significant improvement in blood counts after the surgery. Hemoglobin was 12.5 g/dl, total WBC count was 6,400 cells/cumm and total platelet count was 1.4 lac/cumm. With no further complications, the patient was discharged on the fourth post-operative day.

DISCUSSION

A spleen measuring 400-500 g shows splenomegaly, whereas a spleen weighing more than 600 g is referred to as massive splenomegaly and one weighing more than 1600 g is referred as supramassive splenomegaly.^{3,6,7}

In splenomegaly, hypersplenism develops when a higher proportion of total blood cells are retained for destruction at once, resulting in pancytopenia (anemia, leukopenia, thrombocytopenia). Cirrhosis, infection (malaria, tuberculosis, HIV), autoimmune disorders, leukemia, lymphoma, and hemolytic anemia are among the common causes.⁸

Some patients with splenomegaly are identified with idiopathic splenomegaly after a comprehensive evaluation reveals an uncertain explanation.⁹

At least 2 weeks before or within 2 weeks after intervention, the 23-valent pneumococcal polysaccharide (PN23), the conjugated meningococcal C (MCC), and the Hemophilus influenza type B (Hib) vaccines are indicated in splenectomized patients.⁸

Treatment for hypersplenism varies on its severity. In extreme situations, splenectomy is favored for the majority of hematological reasons and has been selectively employed for its benefits in enhancing survival and cytopenia. Laparoscopic splenectomy is typically preferable over open splenectomy.⁸

Laparoscopic splenectomy is more difficult in individuals with known CHD who are categorized as minor/major/severe based on CHD severity.⁵

Minor

Cardiac condition with/without medication and maintenance with no symptoms, and repair of congenital heart defect with normal cardiovascular function and no medication.

Major

Repair of CHD with residual hemodynamic abnormality with/without medication.

Severe

Uncorrected cyanotic heart disease, patients with pulmonary hypertension, patients with ventricular dysfunction requiring medication, and listed for heart transplantation.

Numerous studies have shown that intraabdominal laparoscopic surgery is safer than open surgery, with lower morbidity and mortality in patients with moderate CHD, but not in those with severe CHD.¹⁰

Pneumoperitoneal pressure has a crucial role in the success of Laparoscopic procedures with fewer complications, particularly in the treatment of CHD.¹¹ The rate of insufflation and intraperitoneal pressure must be regulated with the assistance of an anesthesiologist.

A sudden increase in intraabdominal pressure causes hemodynamic instability, particularly in patients with preexisting heart conditions.¹² IAP more than 12 mm Hg is considered intraabdominal hypertension, which has detrimental effects on the cardiorespiratory system, primarily diaphragmatic splinting and CO₂-induced hypercarbia. Therefore, the lowest feasible IAP must be maintained.¹³

CONCLUSION

In case of supramassive splenomegaly, splenic artery embolization prior to surgery, can greatly help in reducing the risk of intraoperative bleed. Laparoscopic splenectomy in cardiac patients under low pressure (8 to 10 mm of Hg for CO₂ pneumoperitoneum) is feasible and beneficial even in cases of supramassive splenomegaly. Prior vaccinations effectively reduce the risk of post-operative infections – overwhelming post splenectomy infections (OPSI). Splenectomy is the treatment of choice for hypersplenism with pancytopenia as it corrects the altered blood markers in the immediate post-operative period itself, and it not only reduces bleeding complications but also helps in rapid post-operative recovery. Further follow-up to check for platelet count and splenic vein and portal vein thrombosis needs to be done on a regular basis.

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