Torsion of Enlarged Wandering Spleen in Child with Gaucher's Disease

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ABSTRACT

Background: Wandering spleen is a rare condition characterized by malposition of spleen due to laxity or absence of supporting ligaments. Gaucher disease is high risk factor for torsion and trauma of migrant spleen. Case presentation: Six-year's old male patient presented to our institution for emergency treatment with acute abdomen, hemodynamic instability with palpable tender mass in the abdomen, the diagnosis was settled as twisted WS and splenectomy was done. Conclusion: clinical diagnosis of wandering spleen torsion is rare particularly in the pediatric age-group makes it an enigma for the surgical world.

Keywords: Wandering spleen, Gaucher's disease, Acute abdomen, Splenectomy

1. INTRODUCTION

The spleen is a mesodermal in origin from dorsal Mesogastrium develops during 5th week of pregnancy, situated in posterior part of left hypochondrium(1,2). It is fixed in its site by three main ligaments, gastro splenic, lienorenal and splenocolic ligament, and two variably present splenoomental and splenophrenic ligaments(1).

Elongation or maldevelopment of these suspensory ligaments lead to hypermobility of spleen with liability for torsion, infarction that necessitates splenectomy(3). Association of Gaucher's disease increase risk for torsion, here we reported a case of enlarged twisted spleen in a child with Gaucher's disease and splenectomy was performed.

Gaucher disease (GD) is an autosomal recessive storage disorder caused by pathogenic variants in the gene GBA resulting in deficiency of the enzyme glucocerebrosidase glucosylceramidase and pathogenic accumulation of the substrate glucocerebroside glucosylceramide(4,5).

2. CASE REPORT

A male patient, 6 years old presented with acute abdomen with hemodynamic instability and resuscitation was done. The Child had been diagnosed as Gaucher's disease type 1 of sixth months ago, with massive splenomegaly. Patient developed sudden attack of severe abdominal pain and vomiting of 12 hours, on examination of the patient, was very pallor and lethargic, abdominal examination revealed generalized abdominal tenderness maximum in peri umbilical and left lumber regions with palpable, tender and firm mass occupying umbilical, left lumber and left iliac regions.
An array of investigations in form of chest and abdominal plain X-rays as a routine imaging, abdominal US and Doppler US done, moderate enlarged spleen occupying umbilical region with suspecting of ischemic changes, Contrast-enhanced multi-slice CT abdomen which revealed, ischemic WS. Laboratory investigations of patients were:

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<th>K</th>
<th>Na</th>
<th>Creat.</th>
<th>Urea</th>
<th>PT</th>
<th>PC</th>
<th>PLT</th>
<th>WBC</th>
<th>Hct</th>
<th>HB</th>
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<tr>
<td>3.2mEq/l</td>
<td>130mEq/l</td>
<td>.9mmol</td>
<td>20mmol</td>
<td>13minute</td>
<td>68%</td>
<td>140x10</td>
<td>20,000</td>
<td>24</td>
<td>7mg</td>
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Urgent exploration done, delivery of the spleen which was blackish in color, anti-clockwise five times twisted pedicle found, untwist of pedicle done, waiting for 10-15 minutes for regaining vascularity but spleen still blackish and no pulsation in splenic artery. We did splenectomy, exploration for all abdominal organs especially stomach, pancreas and small intestine was free, hemostasis and drain put in bed of removed spleen and anatomical closure of anterior abdominal wall. Twenty four hours post-operative, The patient was good, started oral feeding, drain contain < 50 cc serous collection.

Forty eight hours, patient received triple vaccines (Pneumococcal, Meningococcal and H.Influenza vaccines), after four days patient discharged on penicillin antibiotic and for follow up in outpatient clinic.

Two weeks later patient came to our outpatient clinic; stitches removed and advised to wear abdominal binder 3 months to avoid incisional hernia. Follow up after three months patient was good and gaining weight.
3. DISCUSSION

Torsion of WS is a rare emergency case, as the spleen is at risk of twisting of its pedicle, due to hypermobility, elongation and laxity of supporting ligaments(6,7).

Splenic twisting occur more in two age groups, in children below ten years and in females during childbearing period, usually the condition occur suddenly and patient comes at the emergency unit with acute abdomen and discovered accidently with WS, so clinical diagnosis of twisted spleen is difficult.
Torsion can precipitate by any movement of the body, change intra-abdominal pressure or distension of adjacent organs. TS may result in acute pancreatitis. Defective fusion of the dorsal mesogastrium with the parietal peritoneum results in intraperitoneal location of the Pancreatic tail and leads to ischemic pancreatitis when torsion spleen occurs. Hemorrhagic shock due to sequestration of blood can be the presenting feature of torsion spleen(7).

In our case male child known to splenomegaly, wandering spleen and Gusher's disease type I, presented with acute abdomen with shock, Doppler US and CT scan with I.V contrast diagnosed condition, splenic preservation in form of splenopexy and partial splenectomy were not done due to splenic infarction(8).

4. CONCLUSION

With increase of splenic size especially with laxity of supporting ligaments this may increase liability for splenic pedicle twisting, infarction and splenectomy. Acute abdomen with shock (neurogenic or hypovolemic) may occur especially in children with torsion spleen mostly due to rapid progress of condition which lethal for child, so need proper urgent management.

REFERENCES