NOVEL THERAPY FOR HYPERSPLENISM

DR. THAMBARASI.T,
Post-Graduate

Guide:
Prof.Padmasani.L.N
Dr. Vidya Krishna,
Sri Ramachandra Medical College.
HISTORY:

A 10-year-old developmentally normal boy from Andaman, second born of consanguineous marriage, presented with

- Deterioration in school performance - 6 months
- Difficulty in walking and speech of 5 months duration.
- Stiffness of limbs – progressive in nature

Referred as Suspected Wilson’s Disease

No h/o Jaundice, bleeding manifestations, blood transfusion, recurrent infections.
O/E:
- Vitals normal
- Pallor +
- KAYSER-FLEISCHER RING +
- CVS – S1,S2+
- RS-B/L AE+
- P/A :soft
  - Spleen 11cm below LCM, smooth surface, rounded edges, regular margins.
- CNS: GCS 15/15
  - Dystonia +
  - Dysarthria+
CLINICAL PICTURE
CLINICAL DIAGNOSIS:

- NEURO WILSON
- CHRONIC LIVER DISEASE WITH PORTAL HYPERTENSION
INVESTIGATIONS:

- CBC – Pancytopenia,
  Hemoglobin - 9.1 g/dl
  Total counts - 2000 cells /cu.mm
  Platelets 0.45 lakh per cu/mm
  PS-Microcytic hypochromic anemia, anisopoikilocytosis
- PT, PTT, INR - deranged
- LFT, RFT, Sr. Electrolytes – normal
- USG Abdomen – revealed coarse echotexture of liver with features of portal hypertension
INVESTIGATIONS

- 24 hours urinary copper excretion after d-penicillamine challenge - Elevated (1232.51 mcg/dl).
- **MRI OF THE BRAIN** - T1 and T2 hyperintensity in basal ganglia, putamen, globus pallidus and caudate nucleus
- Upper gastrointestinal endoscopy- gastropathy.
- Serum B12 level was normal.
- Viral markers and ANA were negative
MRI BRAIN
FINAL DIAGNOSIS

- WILSON’S DISEASE WITH NEUROLOGICAL INVOLVEMENT
- CIRRHOSIS WITH PORTAL HYPERTENSION
- HYPERSPLENISM WITH PANCYTOPENIA
TREATMENT

Neuro Wilson’s Disease:
- Planned to start on Trientine
- Problems encountered
  - high cost (Rs. 700/day)
  - Not easily available
  Only Options Zinc, d Penicillamine

Hypersplenism:
Hematologist opinion was obtained. Started on Iron supplements and multi vitamins. Advised against splenectomy.
After extensive literature review, it was decided to take up for Partial Splenic Artery Embolisation.
PROCEDURE

- Selective splenic artery embolisation was done under general anesthesia using coils through right femoral access.
- Post procedure angiogram revealed partial occlusion of splenic artery, slowing of splenic circulation and patent gastro-epiploic artery.
POST PROCEDURE COMPLICATION:

- After 24 hours, he developed fever and severe left hypochondriac pain.
- Serum amylase and lipase were elevated.
- CT of the abdomen showed wedge shaped splenic infarct (14 x 9 x 9 cms) with shrunken liver and dilated portal vein (12 mm diameter).
- POST-EMBOLISATION SYNDROME WITH PANCREATITIS - The symptoms subsided over the next 18 days.
CT ABDOMEN
## Treatment Given:

<table>
<thead>
<tr>
<th>No</th>
<th>Issues</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Wilson’s Disease</td>
<td>D-penicillamine, zinc and multi vitamins</td>
</tr>
<tr>
<td>2.</td>
<td>Dystonia</td>
<td>Trihexyphenidyl, Clonazepam, Baclofen, tetrabenazine</td>
</tr>
<tr>
<td>3.</td>
<td>Hypersplenism</td>
<td><strong>Partial Splenic Artery Embolisation</strong></td>
</tr>
<tr>
<td></td>
<td>Prior to the procedure: Penicillin prophylaxis and received pneumococcal and meningococcal vaccines</td>
<td></td>
</tr>
<tr>
<td>4.</td>
<td>Post-Embolisation Syndrome with Pancreatitis</td>
<td>Analgesics, antibiotics and intravenous fluids</td>
</tr>
</tbody>
</table>
FOLLOW UP:

- On follow up, blood counts gradually improved.
  - 10 days after the procedure.
    - Hb- 11.1g/dl
    - Total count - 5200 cell/ cu mm
    - Platelet - 1,35,000 per /cu mm.
- The child is under treatment for Neuro Wilson’s Disease
- During hospital had urosepsis. Treated. Recovered.
- His siblings have been screened for Wilson Disease.
INTRODUCTION:

- Hypersplenism - a common complication of cirrhosis
  - Enlarged spleen with pancytopenia
  - Normal or hypercellular marrow
  - Improvement in cell counts after splenectomy.
- Is due to sequestration of the cells in the red pulp of the enlarged spleen
- Especially a problem in younger children with larger spleens than in the older age group

Treatment-
- Medical management of the underlying liver disease.
- Surgical options include
  - splenectomy
    - Recently partial splenic artery embolisation (PSE).

RISK ASSOCIATED WITH SPLENECTOMY:
- Post-operative morbidity
- Increased risk of portal vein thrombosis
- Infections by encapsulated organisms, Overwhelming Post Splenectomy Infections (O.P.S.I).
- Worsening of the underlying liver function and encephalopathy.

PARTIAL SPLENIC ARTERY EMBOLISATION

- Minimally invasive procedure involves trans-catheter occlusion of the splenic artery and/or its branch vessels.
- Done using gel foam, polyvinyl alcohol particles (PVA), coils.
- Embolisation of about 40-80% of the splenic tissue is a preferred option to manage hypersplenism in cirrhotics.

INDICATIONS

- Cirrhosis with Portal Hypertension and Hypersplenism
- Idiopathic Thrombocytopenic Purpura (ITP)
- Hereditary Spherocytosis
- Thalassemia
- Autoimmune Hemolytic Anemia
- Splenic Trauma/ Hemangioma
ADVANTAGES

- The risk of infections and worsening of liver function is reduced as some functioning splenic tissue and splenic circulation are preserved.
- Unlike splenectomy, PSE decreases the risk of variceal bleed and refractory ascites and improved hepatic function.
- The rise of blood counts occurs within 2 weeks of PSE and is directly proportional to the volume of spleen targeted.

COMPLICATIONS

Post-embolisation syndrome is the commonest complication encountered in 78-100% of the patients

○ Usually begins after 24 to 48 hours and will last for several days.

○ Clinical Features: low grade fever, left hypochondriac pain and anorexia.

○ Treatment: self limited and is managed conservatively with analgesics and intravenous fluids. High grade fever should raise the suspicion of infection.

- Risk of embolisation, being higher if more than 50-70% of the splenic artery is occluded. Embolisation causes tissue ischemia, which produces inflammation and releases cytokines.
- Pancreatitis, though uncommon, has been reported as a complication of PSE.
- Left sided pleural effusion or ascites,
- Splenic abscess

FOLLOW UP

- Routine Computed tomography (CT) abdomen should be done after PSE to see the extent of splenic infarct.
- Infection risk can be reduced by antibiotic prophylaxis in the pre-and post-operative period for upto two weeks
- Vaccinations may not be required.
- Mortality rate is around 0-6%

1. Amin et al-
- 40 cirrhotics treated for hypersplenism by either splenectomy or PSE.
- Hematological indices increased in both the groups within 2 weeks.
- PSE- 20 patients, 1 died of myocardial infarction, 1 developed splenic abscess and 1 developed portal vein thrombus.
- Splenectomy group, 20 pts- 3 patients developed portal vein thrombosis. Overall, longer procedure times and hospitalizations, required more transfusions and post procedural analgesia.

2. K. Ohmagari et al studied thirty patients with portal hypertension and hypersplenism, PSE significantly improved the clinical manifestation of portal hypertensive gastropathy


3. Ron et al - 50 patients underwent SPEs. All embolizations were technically successful. The procedure efficacy was 90%. Side effects included hydrothorax (n=26, 52%), thrombocytosis (n=16, 32%), thrombocytopenia (n=13, 26%), and postembolization syndrome (n=11, 22%).

TAKE HOME MESSAGES:

- PSE - safe and effective option for managing hypersplenism in cirrhotic children
- Advantages - preserved splenic function, reduced risk of encephalopathy
- Complications can be limited by limiting the degree of splenic artery embolised to 50%.
ACKNOWLEDGEMENT

- Prof. Padmasani
- Dr. Vidya Krishna
- Prof. Santhosh Joseph, Interventional Radiologist
- PICU Team
THANK YOU ........