



A RARE CASE OF LIVER CIRRHOSIS AND PORTAL HYPERTENSION WITH HUGE SPLENOMEGALY, HYPERSPLENISM & SPLENIC PSEUDOCYST

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ABSTRACT

Portal hypertension is a common problem in our country. It occurs as a complication to periportal fibrosis that result from schistosomiasis, it a medical condition unless complicated. the complications may manifest as hyperactivity of the spleen and destruction of all blood cell types that result in pancytopenia as in this case. The enlarged spleen may undergo some degenerative changes such as splenic infarction that complicated by pseudocyst as in this case. A case reported of 56 yrs old known case of periportal fibrosis, portal hypertension and liver cirrhosis complicated by huge splenomegaly, hypersplenism and post infarction splenic cyst which develop rapidly over period of 10 days during hospital admission. It's very rare to find all these findings on the same pt and to our knowledge there's no literature reported similar case.

KEYWORDS: Periportal fibrosis, schistosomiasis.

INTRODUCTION

Splenomegaly is the enlargement of the spleen (spleen is an organ of left upper quadrant of the abdomen it's usually fist shape, purple and 4 inches long). splenomegaly is one of the four cardinal signs of hypersplenism which include reduction in the circulating blood cells and compensatory proliferative respons in the bone marrow.

Splenomegaly could be a complication of ppf and liver cirrhosis which is commonly caused by visceral schistosomiasis in Sudan.

Portal hypertension can be a result of periportal fibrosis and liver cirrhosis, which is defined as hepatic venous pressure more than 12 mmHg^[1] and it has many manifestations like esophageal varices.

Sign of splenomegaly may include left upper quadrant abdominal mass or splenic rub. it can be detected on physical examination by castell's sign, traube's space percussion or Nixon's sign and U/S can be used to confirm diagnosis.^[2]

Splenic cysts are rare with around 800 cases reported in the world literature.^[3] Cysts of the spleen can be classified to parasitic and nonparasitic. Non parasitic

cysts can be further classified as true cysts and pseudocysts. Pseudocysts don't have epithelial lining and are usually post traumatic or inflammatory degenerative.^[4]

CASE REPORT

54 yrs old male not known to be diabetic or hypertensive, alcoholic 4glasses /wk, also had history of untreated schistosomiasis at the age of 20 yrs, and was diagnosed 4 yrs ago as a case of liver cirrhosis, portal hypertension and huge Splenomegaly as a result of periportal fibrosis and planed for medical treatment and follow up, but the Patient stops his medication.

3 months ago the Pt came complaining of fatigability and shortness of breath, diagnosed as anemia and referred for further evaluation that showed the cause of anemia is hypersplenism as a complication of portal hypertension.

After stabilization of Patient condition a Surgical Consultation to our unit done and decision of elective splenectomy was made after detailed Clinical Examination and Investigations and child's criteria was calculated.

On Clinical Examination

The Patient was unwell, cachexic (wt: 56 kg), pale but not jaundiced. all systems were normal on examination.

On abdominal examination

Abdomen was distended with full flank and central flat umbilicus.

- No dilated veins, no area of guarding, rigidity or tenderness.
- No stigmata of chronic liver disease (clubbing, gynecomastia, caput medusa, spider neva), or signs of liver decompensation
- Liver 15 cm below costal margin, nodular rough surface, and liver span 28.
- Spleen 33cm below costal margin extending to right iliac fossa.
- Moderate ascitis, positive shifting dullness.

Investigation

The following investigations requested for more evaluation:

- CT scan that showed (huge Splenomegaly, liver cirrhosis and moderate ascitis).
- Abdominal U/S that showed (portal vien about 2.5 cm, huge Splenomegaly, liver cirrhosis and moderate ascitis).
- Upper GI endoscopy that showed no oesophageal varices but there's gastropathy.
- CBC showed pancytopenia of all cell lines, RFT and electrolytes (normal), LFT (normal) and chest x-ray (elevated left hemidiaphragm).
- Bone marrow biopsy showed hypercellularity of all cell lines.

After the investigations and as a part of the Patient preparations to surgery he received vaccinations against: H.influenze, Pneumococcal and Meningococcal vaccines also anti-malarial medication had been given according to protocol.

During the period of admission the Patient developed

severe pain at the site of the enlarged spleen continued for 6 hours, after 2 days from this complain the Patient developed cystic mass that increased progressively in size over period of 10 days at the site of pain.

This condition diagnosed as post infarction splenic cyst and abdominal U/S is requested and shows huge cystic lesion on top of the Splenomegaly about 20×25×19 cm.

This mass is consisted with post infarction splenic pseudocyst which is very rare and not reported throughout the world.

It's not rare to find Patient with liver cirrhosis and portal hypertension in our society but it's rare to find it complicated by hypersplenism and post infarction pseudocyst.

Child – puph was calculated for this patient and accordingly our patient is child “B”.

At Surgery

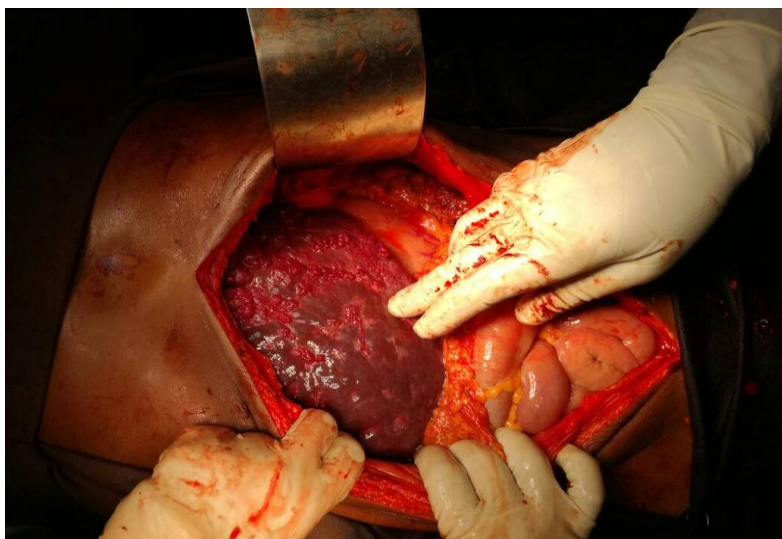
Longitudinal midline incision done, spleen was extending to right iliac fossa (wt= 8.9kg and long 39.5×20.9cm) with large splenic cyst (30×25×20cm) at the lower border of the spleen, also a large diameter of splenic vein (3.1 cm) noticed, with normal splenic artery diameter, liver was enlarged with rough nodular surface.

Specimen was send for histopathology and fluid from cyst for culture, & cytology result (heamorrhagic aspirate contain old red blood cell and cell debris).

Spleen normal in histology.

There's large pseudocyst with epithial lining on a top of infracted area of spleen.

Post operatively it was uneventfully and the Patient discharge at 4th postoperative day.





Spleen & splenic cyst before removal (inner view)



Spleen & splenic cyst before removal (external view)



Spleen & splenic cyst after removal (inner view)



Hugely dilated splenic vein with normal splenic



Hugely dilated splenic vein insitu before cutting & ligation, hold between surgeon index & thumb



Hugely dilated splenic vein after splenectomy

DISCUSSION

Portal hypertension due to liver cirrhosis and periportal fibrosis is a common clinical problem, splenomegaly is important sign for diagnosis of portal hypertension.

Portal HTN also may cause huge splenomegaly that cause pressure to bowel and stomach and left hypochondrial pain but most serious complication of o portal HTN and splenomegaly is hypersplenism.

Hypersplenism may affect all cell lines as in our case or may affect a single line.

Pateint with hypersplenism susceptible to anemic heart failure, bleeding tendancy and recurrent infections.

We reported another complication of huge splenomegaly;

splenic cyst which is a very rare clinical problem that can be primary (true cyst) have epithelial lining or secondary (pseudocyst) have no epithelial lining.^[5]

Primary cyst can be parasitic (most commonly echinococcal) or Non Parasitic(simplecyst, dermoid, epidermoid and neoplastic cyst).^[6]

Splenic pseudocysts account for about three quartant of all non parasitic splenic cysts. Usually result from splenic parnchymal trauma or subcapsular heamatoma. Some times arise after splenic infarction, degenartive changes and infections (malaria, infectious mononucleosis and tuberculosis).^[7,8,9,10]

What happend in this case is post infarction splenic pseudocyst which is a rare condition.

Juma et al, report a non-traumatic splenic cyst that present with left upper quadrant pain and measured 16×10. Cm but their pt was quite well and not acase of liver cirrhosis, periportal fibrosis, portal hypertension or splenomegaly, and the cyst was of unknown cause.

In this case the patient is a known case of liver cirrhosis, periportal fibrosis, portal hypertension and huge splenomegaly and his cyst developed after infarction of the spleen and the cyst measured 30×25×20 cm.

Kiran et al, reported a large pseudocyst measuring 27×20 cm also the cyst of unknown cause.^[11]

»Valentina et al, reported a large splenic cyst 20×13 ×21 cm, it was aprimary cyst and in adolescent. Which is inconsistent with our finding.

Largest splenic cyst reported world wide measuring 30×30×30 cm but it,s a primary cyst^[12], not as in this case of a largest pseudocyst on top of infarcted area.

Most of pseudocysts is unilocular (80%) and it varies in size (1-16 cm).

Concerning clinical presentation most of the cysts are a symptomatic (30-60%) and diagnosed incidentally.^[13,14]

Aspiration of fluid from cyst donor differentiate true from pseudocyst or subclassify the conginital cyst.^[15]

Diagnosis of pseudocyst based on radiological (U/S, CT, MRI) histopathological, haematological, biochemical, bacteriological and serological studies. To rule out other causes of splenic cyst.

U/S useful for determination of size, site, nature and echogenicity of the cyst.

CT give more details, architecture of spleen, location and its relation to the surrounding structure. MRI used when US and CT scan are inconclusive.^[11] Different surgical modality can be employed it,s include : total splenectomy, partial splenectomy partial cystectomy, morsuplazation, and percutaneous drainage^[16] our case already planed for total splenectomy and cyst developed during the period of preparation to surgery.

There is no literature reported hugely dilated splenic vein as in this case it measured 3,1 cm.

Liver cirrhosis, portal HTN, huge splenomegaly (39.5×20.9cm), hypersplenism, a large pseudocyst on top of infarcted spleen(30×25×20), hugely dilated splenic vein all this findings in one patient; after throughly review to literature there is no reported similar condition.

So we believe that we successfully operated the largest spleen and largest splenic pseudocyst up to date.

CONCLUSION

Liver cirrhosis, ppf and portal HTN is common in our country, hypersplenism usually complicate portal hypertension and extremely huge spleen prone to infarction and may undergo a cyst on top of infarction.

This can occure suddenly and progressively, so patient with portal HTN should a close follow up to detect and treat this complication as early as possible.

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