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Case Report

Budd Chiari syndrome due to inferior vena cava web complicating pregnancy in patient with bad obstetric history: an interesting case

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ABSTRACT

Presenting an unusual case of 27 years old female who presented at 18 weeks of pregnancy with second trimester bleeding per vaginum. Patient had history of recurrent abortions on examination was found to have hypertension and thrombocytopenia. Usg done revealed severe oligohydramnios. Patient was managed conservatively but aborted spontaneously at 22 weeks of gestation. Post-abortionl on day 2 patient developed abdominal distension and liver function tests were found to be deranged. USG and CT abdomen and pelvis was done, which revealed Budd chiari syndrome due to inferior vena cava (IVC) web. This extremely rare condition is characterized by obstruction of inferior vena cava by membrane or fibrous band. This condition is diagnosed by radiological techniques which in our patient revealed classical findings of caudate lobe hypertrophy, non-visualization of hepatic veins, moderate hepatomegaly and spleenomegaly and multiple collaterals. Esophagogastroduodenoscopy done which revealed large varieces for which endoscopic variceal ligation was done. IVCgram and IVC plasty was done by interventional radiology department 6 weeks after abortion. The aim of this case report is to highlight an extremely rare cause of Budd Chiari syndrome and IVC web in patient with recurrent abortion with spleenomegly leading to thrombocytopenia. It is important to rule out other differential diagnosis in these patients like APLA, ITP.

Keywords: Budd Chiari syndrome, Hepatosplenomegaly, IVC web, Portal hypertension, Recurrent pregnancy loss, Thrombocytopenia

INTRODUCTION

Budd chiari syndrome is an uncommon condition caused by the obstruction of hepatic outflow tract.[1] It commonly implies thrombosis of hepatic vein and or intra hepatic or supra hepatic IVC. Presentation may vary from completely asymptomatic to fulminant liver failure.1 Budd chiari syndrome is an example of post-sinusidal portal hypertension.[1] Budd chiari syndrome is a rare and life threatening disorder.[2] This veno occlusive disease occurs due to various causes like myeloproliferative disease, hypercoagulable states like leiden factor 5 mutation, antithrombin 3 deficiency, antiphospholipid antibody syndrome, protein C deficiency, protein deficiency, pregnancy, oral contraceptives, membranous web in IVC and idiopathic causes. There is very little literature available about its occurrence with pregnancy. Budd chiari syndrome during pregnancy has rarely been reported.[3] It may present as acute disease with severe upper abdominal pain, jaundice, hepatomegaly, ascites,variceal bleeding. Chronic disease presents as abdominal pain of long duration, massive ascites, hepatomegaly, cirrhosis. diagnosis is done with imaging modalities like doppler ultrasonography, CT scan, MRI,

venography, arteriography. Early diagnosis is required for appropriate treatment.[4] Proper clinical history and imaging are essential for definitive diagnosis.[4] Management includes medical treatment with anticoagulants and thrombolytics. Surgical options are angioplasty, TIPS, shunts, liver transplant. The main goal of treatment is to alleviate hepatic congestion, thereby improving hepatocyte function and allowing resolution of portal hypertension.[5] Several articles and case reports on budd chiari syndrome conclude that hypercoagulable states are its main cause in most patients. The important etiologic factors are related to hypercoagulability of blood.[6] This case report throws light on a rare case of Budd chiari syndrome which is IVC web that we authors came across while treating a pregnant lady with recurrent abortions, thrombocytopenia and spleenomegaly. In post abortal period when she developed abdominal distension imaging modalities were used to detect the cause and the CT scan suggested that probable explanation to her symptoms was a web in IVC that might be the cause for Budd chiari syndrome that she had developed.

CASE REPORT

History of the case

Mrs. ABC 27 years old female patient G3A2 with 19.2 weeks by dates, 18.1 week by USG came with chief complaints of spotting PV and fever since 2 days and platelet count of 71,000.

- Known case of chronic hypertension on
- Tablet labetalol 100 mg twice a day
- Diagnosed as hypothyroid after admission
- No history of heavy weightlifting/Trauma
- No history of recent intercourse
- No history of pain in abdomen or leaking per vaginum
- No history of DM/TB/Asthma/epilepsy.

Obstetric history

- Gravida 1 spontaneous abortion in 2016 at 4th month of pregnancy, check curettage done
- Gravida 2 spontaneous abortion in 2017 at 3rd month of pregnancy, check curettage done
- Gravida 3 present pregnancy.

On examination

- General condition fair
- Afebrile
- Pulse: 90|min
- BP: 130|90 mmHg
- CUS: heart sounds normal
- RS: clear
- PA: uterus 18 weeks, fetal heart sounds present, 140 beats per minute, relaxed
- PS: cervix healthy, vagina healthy, altered discharge present
- PV: uterus 18 weeks, bilateral fornices free, no active bleeding

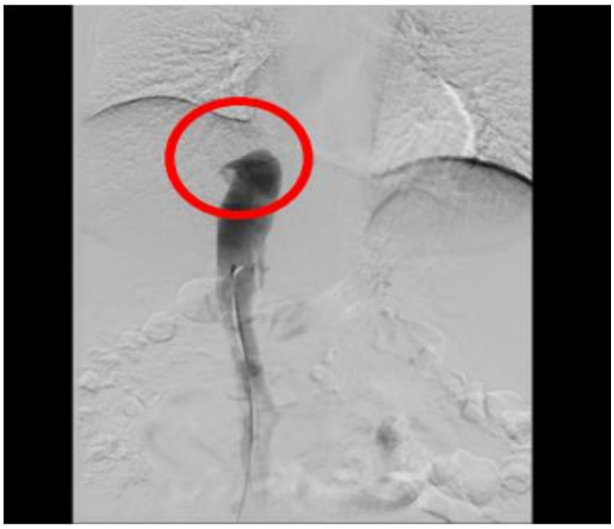

Figure 1: IVC gram taken before intervention: non-contrast area (highlighted with red circle) suggestive of block in the inferior vena cava.

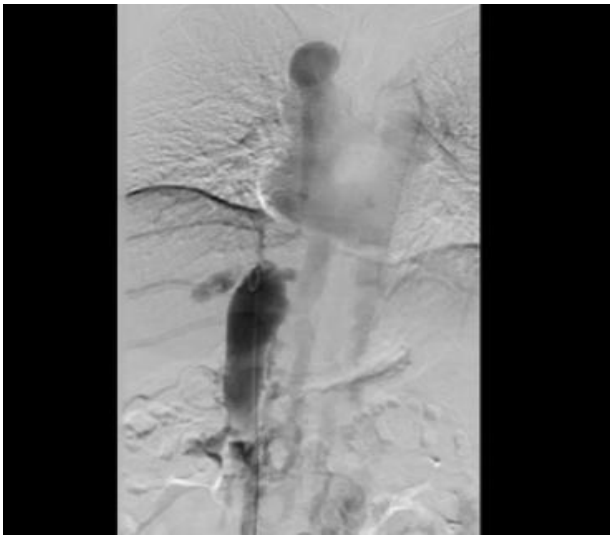

Figure 2: IVC gram taken before intervention: the developed collaterals as compensatory mechanism by body as the interior vena cava is not patent. (The dark black appearance is of inferior vena cava and to its left are the light black appearance or light black parallel lines suggestive of collaterals).

USG Obstetrics

- Single live intrauterine gestation of 18.2 weeks
- Cephalic
- Placenta Fundo-posterior
- A large 10.8x4x7.8 cm sized heterogeneous lesion with few hypoechoic to anechoic areas within noted

on fetal side of placenta with no internal vascularity within suggestive of preplacental hematoma.
- AFI - 6.1 cm.

ANC profile

- Hb/platelets: 8.1/71000
- Blood Group: B+ve
- HIV: NR
- HBsAg: NR
- VDRL: NR
- FBS / PLBS: 90|128
- SrTSH: 22.95.

Other investigations

- LFT: Bilirubin: 0.9/sgot: 36/sgpt: 16,
- RFT: Bun: 7/creatinine: 0.6, PTINR 1.14
- ANA negative, anti DS DNA negative
- ACL IgG, IgM negative, lupus anticoagulant negative
- HB electrophoresis - No abnormal haemoglobin
- Dengue PCR and leptoPCR negative
- Blood cultures no growth
- High vaginal swab - no growth

Management

Patient was managed conservatively in antenatal ward.

- Injection Proluton 250 mg IM weekly
- Tablet susten 400 mg twice a day
- Tablet labetalol 100 mg twice a day
- Tablet thyronorm 150 µgm OD as per endo medicine reference taken for raised TSH level.
- IV antibiotics were started as patient had repeated fever spikes (medicine reference was taken; cultures and iv antibiotics were advised by them). In Spite of conservative management patient developed severe oligohydramnios and aborted spontaneously and completely on 28/8/18 at 24.5 weeks of gestation
- Unit platelet was transfused as Platelet counts post abortion was 40,000.

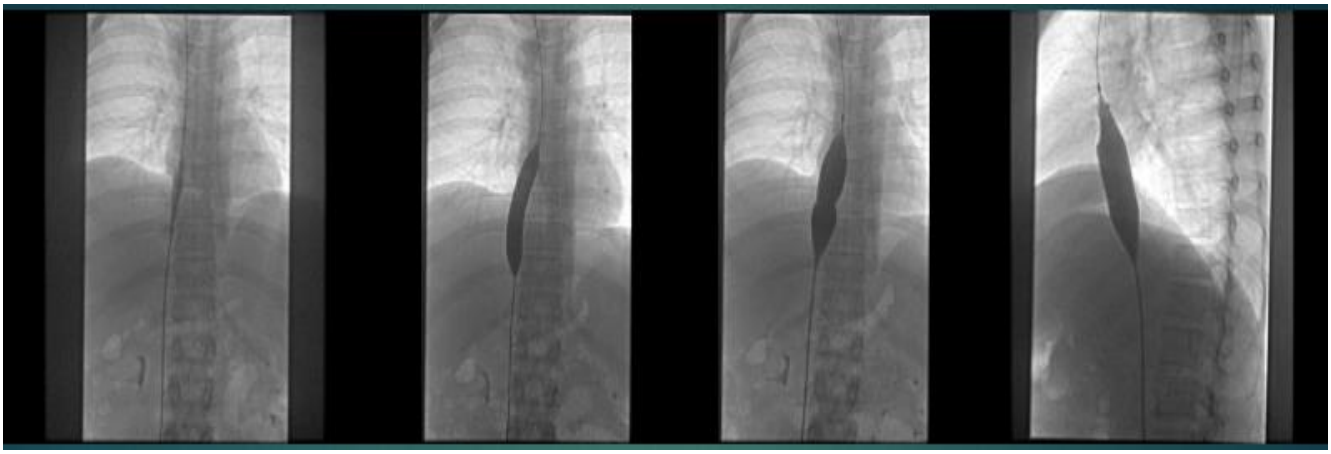

Figure 3: X-ray taken while balloon dilation was being done: Images showing stenting being done with serial balloon dilation (in the images it can be seen that the balloon that is seen as dark black, is dilated gradually to establish the patency of the blocked vessel).

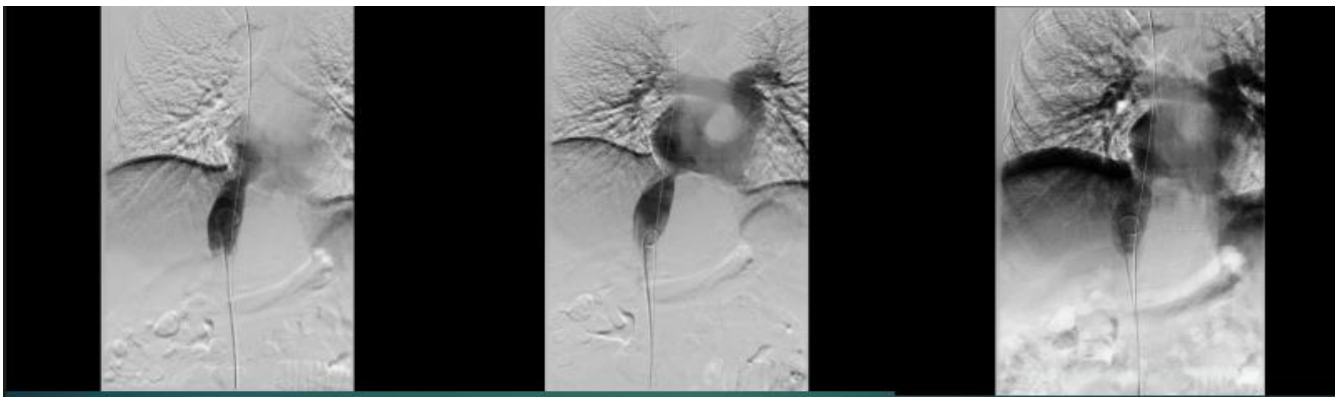

Figure 4: IVC gram taken after radiological intervention (IVC plasty): post intervention image showing patent inferior vena cava with normal blood flow (as the contrast shows patent vessel).

Post abortion patient developed abdominal distention

AG: increased by 5 cms on day 2 of abortion, LFT: total bili 2.5 SGOT/SGPT: 27/28 were also found to deranged, so USG SCAN was done that revealed following findings:

- Liver-coarse echotexture size 13.5 cm caudate lobe hypertrophy seen
- Spleen normal
- Uterus bulky (postpartum status)
- No internal vascularity, ET 12 mm
- Few splenic hilar and lienorenal collaterals and large abdominal wall collateral
- Terminal portion of left hepatic and middle hepatic vein not visualized with few venovenous collaterals.

Features suggestive of Budd Chiari syndrome CT abdomen and pelvis findings

- Mild hepatomegaly, irregularity with caudate lobe hypertrophy suggestive of chronic liver parenchymal disease
- Moderate splenomegaly
- A 2 mm thick linear transverse filling defect is noted involving suprahepatic segment of IVC with resultant mild to moderate luminal narrowing possibly suggestive of IVC web
- RHV is seen with a large intrahepatic venovenous collateral ms approx. 1.6 cm which is draining into IVC. MHV is not well appreciated? joining RHV. LHV is not draining into IVC, instead it is seen joining RHV through another collateral over surface seen
- Multiple intrahepatic veno-venous collaterals seen
- Dilated portal vein and splenic vein seen
- Multiple dilated tortuous collaterals in oesophageal region(submucosal), perigastric region, (few of these gastric collaterals are intraluminal along the lesser curvature of stomach), splenic hilar, lineorenal, along bilateral adrenals. Anterior abdominal wall, in mesentery seen.

The mentioed features suggestive of IVC web with Budd chiari syndrome with features of portal hypertension

- Gastro medicine reference taken
- Oesophagogastroduodenoscopy (OGD) done. (Large esophageal varices seen)
- EVL (Endoscopic variceal ligation) done
- IVC gram was done that showed block in IVC (Figure 1) and development of collaterals by the body in response to the block (Figure 2). This was followed by IVC plasty by serial dilatation with balloon (Figure 3) by intervention radiologist. IVC plasty was successful and the IVC gram taken after the intervention showed patent IVC vessel and reestablishment of normal blood flow in the initially blocked vessel (Figure 4).

DISCUSSION

Budd chiari syndrome is a veno occlusive disease that is rare but can be diagnosed with current imaging modalities and timely management can help in decreasing its complications. Budd chiari syndrome mainly affects women of child bearing age.[7] Imaging plays a crucial role in early detection and assessment of extent of disease in Budd chiari syndrome.[8] The key imaging finding in Budd chiari syndrome are occlusion of the hepatic veins, IVC or both; caudate love enlargement and presence of intra hepatic collateral vessels and hyper vascular nodules.[9] The selective use of anti-coagulation, vascular stents, trans jugular intra hepatic porto systemic shunt and liver transplant has resulted in increase in survival.[10] In our patient recurrent abortions, thrombocytopenia and spleenomegaly was initially managed conservatively. Blood investigations were done to rule out conditions like ITS and antiphospholipid antibody syndrome. In the post abortal period the sudden presentation of abdominal distention finally lead to diagnosis of Budd chiari syndrome due to IVC web and patient was managed with the help of interventional radiologist.

CONCLUSION

The aim of this case presentation is to highlight an extremely rare cause of Budd Chiari syndrome with IVC web in a patient with a history of recurrent pregnancy loss, thrombocytopenia, hepatosplenomegaly, and post-abortion abdominal distention.

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