

## Osler Weber Rendu Syndrome in Pregnancy

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## Case History

- 29 years old, G4 P3 L2, presented at 29 weeks with right hypochondrial pain associated with recurrent episodes of nasal bleed

## Further History

- Asymptomatic cholestasis
- Family history of recurrent epistaxis ( mother)
- 2 Full Term-SVD; uncomplicated
- 3<sup>rd</sup> pregnancy complicated with recurrent epistaxis and haematemesis which subsequently complicated by IUFD, DIC, sepsis, congestive liver and portal cavernoma at 32 weeks. Required multiple transfusions before delivery

## Examination Findings

- Mild pallor, tachycardia (PR 100bpm), normotensive; wide pulse pressure
- Prominent Systolic murmur in all areas of precordium
- Tender Right hypochondrium
- Liver not palpable as it was obscured by gravid uterus

## Investigations

Haematological report	Hb (8.7g/dl) , PLT (2.39 lakhs/cumm), WBC(8,400/cumm)
Coagulation profile	PT (18.6sec~ control 13.5sec), APTT (35.7sec~control 28sec), INR 1.56
Biochemistry Report	Sodium(136mEq/L),Potassium (3.9mEq/L), LDH (196U/L) amylase( 34.0U/L)
	Total Bilirubin (1.0mg/dl), Direct Bilirubin (0.2mg/dl) ALT (34U/L), ALP (183U/L) Total protein (5.8gm/dl)
	TSH 1.46uU/mL, FT3(2.4uU/mL) / Se Ferritin (99.4ng/mL)

## Investigations

- ECG - Left Ventricular overload
- Abdominal USG
  - Congestive liver
  - Dilated portal vein and intra-hepatic portal venous radicles, superior mesenteric vein and proximal splenic vein.
- Impression : chronic spleno-portal axis thrombosis with cavernoma formation

### Echo-cardiography

- mildly dilated chambers evidence of hyperdynamic circulation

### Upper Gastro-Intestinal Endoscopy

- Active bleeding from nasopharynx with normal study in other parts.

### Diagnostic Nasal Endoscopy

- Multiple mucosal bleeding points on right side and small bleeding point at nasopharynx

### Differential Diagnosis of Hyperdynamic Circulation and High Output Failure

- Osler weber rendu syndrome
- Portal cavernoma with periportal arterio venous communication +/- Budd chiari syndrome

### Treatment

- Managed conservatively:
- Multidisciplinary care involving:
  - Physicians / Anaesthetists / Obstetricians
  - Radiologists / ENT specialist
  - Gastroenterologists (medical and surgical)
  - Pulmonologist, Cardiologist, Nutritionists etc

### Treatment

- Montelukast sodium
- Nasivion nasal drops
- Mupirocin ointment for epistaxis
- Haematinics
- Pain relief PRN with fentanyl, pethidine, tramadol; other supportive care

### Discharge

- At request after 7 days of admission
  - Haemoglobin at discharge: 8.7g/dl

### Plan of Management

- Readmission
- In-patient monitoring till 34 weeks
- Delivery after 34 weeks
- CT chest and MRI abdomen for further delineation of vascular abnormalities in chest and abdomen

### Re-admission

- Readmitted 6 days later at 31+weeks with:
  - Severe right upper abdominal pain and an episode of epistaxis
  - Blood pressure was 110/60mmHg, PR 110 bpm
  - Had bilateral pedal oedema.

### Clinical Diagnosis

- Clinically diagnosed to be complicated with high output heart failure
- Admitted to Intensive Care unit; treatment for heart failure was started

### Investigations

- Haemoglobin : 8.7g/dl
- Platelet count : 2.43 lakhs/cumm
- Total WBC : 9,300/cumm
- PT : Test: 16sec ~ control: 13.5sec
- APTT : Test: 36.7sec ~ control: 28sec  
INR 1.24
- Other biochemical parameters were normal

### CT Scan Chest with Contrast

- Multiple pulmonary AV malformation area noted at both lungs
- Largest at right upper lobe measuring 4.4 x 2.6cm
- AV malformation at right middle lobe is measuring 3.3 x 2.5cm
- Left lower lobe shows small AV malformation measuring 2.5 x 2.1cm

### CT scan of Upper Abdomen

- Enlarged liver with grossly altered attenuation at liver parenchyma with different enhancement.
- Grossly dilated intra-hepatic, hepatic venous radicals.
- Multiple collaterals and AV shunts are seen at porta-hepatis and along the walls of gall bladder.
- Solitary large calculus measuring 1.7cm seen in gall bladder

### MRI Upper Abdomen

- Liver is enlarged in size with no focal lesion seen.
- Multiple dilated flow void signal intensity lesions noted around the porta-hepatis, around the wall of gall bladder, at the retroperitoneum and in the small bowel mesentry

### Plan for Delivery

- In-view of her condition (worsening high output cardiac failure with recurrent right hypochondrial pain affecting her nutrition as well), she and her family were counselled and a decision to deliver was made at 32 weeks gestation

### Labour Analgesia

- The decision for epidural analgesia made cautiously after considering every angle of safety issue in methods of analgesia. High lumbar epidural with open ended catheter was inserted using continuous saline flush technique to minimise blood trap.

### Labour / Delivery details

- Labour was induced with PGE1
- But after several hours caesarean section was done under epidural analgesia at patient's request. Tubal ligation was performed. Delivered baby girl of 1.66kg

### Postoperative Period

- She received 2 units of PRBC transfusion to improve her hematocrit in view of persisting right hypochondrial pain.
- Post operatively her recovery period was complicated with wound infection which required antibiotic and drainage.
- She otherwise recovered satisfactorily

### Plan of Management

- To be reviewed by intervention radiologist at a higher centre to tackle the AVMs causing hyperdynamic circulation.