AN INTERESTING CASE OF RESPIRATORY DISTRESS

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• 11 month old female infant 1st born to parents of NC marriage referred from Kolkatta

• H/O:

Acute onset of respiratory distress

2 episodes in 1 month
• HOPI:
• Apparently Normal 1 month back
• Developed URI asso with breathlessness
• Admitted & treated as bronchiolitis in Kolkatta
• Inv : Normal except for cardiomegaly in CXR
• D/S with adv to follow up after 1 week
• Within 5 days after the D/S child again developed severe respiratory distress asso with profuse sweating, puffiness of face, edema of leg

• Not asso with Bluish discolouration

• Admitted & investigated at Kolkatta

• Found to had PHT with CCF

• Treated with Sildenafil, Aldactone, Lasix & Digoxin

• Referred to RH PER for further evaluation
• PAST HISTORY:

Remarkably Normal child
Well Thriving
No H/O CHD, RRTI
NO H/O Seizures
No significant hospitalisation
H/O dyspnea with feeds on & off, minimal sweating not asso with feeds not warranting any hospitalisation for the past 1 month
• No H/O young stroke, liver disease, hematological disease in the family
• No BOH in mother
• Uneventful Antenatal Period
• FT / LSCS / B.wt – 3 kg
• Neonatal period:
  Treated for neonatal sepsis
  No H/O DIC, NPF
• O/E:

Afebrile
No pallor, icterus, cyanosis, clubbing.
Peri orbital edema +
Minimal abd distention +
B/L pedel edema +
CVS / RS / CNS : NAD
P/A : Liver 3 cm
Liver Span : 7cm
• INV:
  • Hb : 12.1
  • PCV : 38%
  • Plt : 2,29,000
  • TC : 13,000
  • DC : P-17 % ; E-1% ; L-74% ; M-8 %
  • PS : Normal Study
  • LFT / RFT / Electrolytes : WNL
- Cardiolipin Ab IgG: 28 GPL units/ml (Positive- >10 )
- Cardiolipin Ab IgM:10 units/ml (equivocal 10-15)
- Lupus Anticoagulants : 68.5 ( Inc )
- Anti thrombin III activity : 51% (dec)(72-134%)
- Protein C : 18 (dec ) (31-112%)
- Protein S : 15 (dec ) (29-162%)
- Homocysteine : 15.8 ( N )
- APTT : Prolonged
• CXR : Cardiomegaly

• ECHO:
  Dilated RA / RV
  Increased PAP
  RPA: Continuous flow from MPA with shagginess in the branches
  LPA: Could not be made out
• CT ANGIO:
  Thrombotic lesion in both Pulm. Vessels
  No underlying structural anomaly found

• DOPPLER BOTH LL : Normal
FINAL DIAGNOSIS

ANTIPHOSPHOLIPID ANTIBODY SYNDROME
COURSE

- Started on Heparin infusion
- Posted for Thrombectomy
- Went for cardiac arrest and collapsed
- Further investigation and screening of the family members could not be done as they were from outside Chennai and not willing for further investigations
DISCUSSION
Antiphospholipid syndrome is also referred to as Hughes syndrome after the rheumatologist Dr. Graham R.V. Hughes

Antiphospholipid syndrome
- Disorder of coagulation.
- Thrombosis in both arteries and veins
- Pregnancy-related complications such as miscarriage, stillbirth, preterm delivery, preeclampsia
TYPES

• **Primary APS:**
  - APS occurs in the absence of any other related disease.

• **Secondary APS:**
  - Seen in conjunction with other autoimmune diseases
  - APS coexists with SLE, Malignancy

• **Catastrophic APS:**
  - APS leads to rapid organ failure due to generalised thrombosis and a high risk of death in short duration
• Autoimmune production of antibodies against phospholipid substance.
• Characterised by antibodies against cardiolipin, and β2 glycoprotein I
• β2-glycoprotein I is the predominant target of autoimmune antiphospholipid antibodies
• other phospholipid-binding proteins include prothrombin, protein C, protein S and annexin V
Putative "second" hit
- trauma
- infection
- nonimmune procoagulant factors

Anti-lamin B1 antibody exerts protective effect

ANTIPHOSPHOLIPID SYNDROME

APA binding to β2-glycoprotein, prothrombin, proteins C and S, and annexin V interferes with coagulation cascade

- Protein C activation
- Antithrombin III activity
- Annexin V binding
- Fibrinolysis
- Tissue factor activity
International Consensus Statement - Update of the Classification Criteria for Definite Antiphospholipid Antibody Syndrome

A patient must meet at least one clinical and one laboratory criterion for a diagnosis of antiphospholipid antibody syndrome.
• **Updated clinical criteria**

• **Vascular thrombosis** - Arterial, venous, or small vessel thrombosis in any tissue or organ confirmed by imaging studies, Doppler studies, or histopathology (without significant vessel wall inflammation)

• **Pregnancy morbidity**:
  
  – Fetal deaths at more than 10 weeks’ gestation
  
  – Premature births at less than 34 weeks’ gestation due to severe preeclampsia, eclampsia, or placental insufficiency
  
  – Three or more unexplained consecutive spontaneous abortions at less than 10 weeks’ gestation, excluding maternal anatomic or hormonal abnormalities and paternal and maternal chromosomal abnormalities
• Updated laboratory criteria

• Anticardiolipin (aCL) antibody of Ig G / IgM in medium/high titer measured by a b2-GPI–dependent ELISA.

• Lupus anticoagulant on 2 or more occasions at least 6-12 weeks apart.
  – Prolonged phospholipid-dependent coagulation (eg, aPTT, Kaolin clotting time [KCT], dilute Russell viper venom test, dilute PT)
  – Failure to correct the prolonged coagulation time by a mix with platelet poor plasma (PPP)
  – Shortening or correction of the prolonged coagulation time with excess phospholipid
  – Exclusion of other coagulopathies
CLINICAL MANIFESTATIONS

- The most common – THROMBOSIS
- Venous thrombosis
  - Lower limb, Lungs
- Arterial thrombosis
  - Brain in up to 50% - TIA or strokes.

Others:
- Heart (25%), causing coronary occlusion
- Eye, kidney and peripheral arteries (25%).
• In pregnant women:
  - Miscarriage (prior to 20 week of gestation)
  - Pre-eclampsia
  - Placental infarctions
  - Stillbirth
• Mental and/or development retardation in the newborn, due to an aPL-induced inhibition of trophoblast differentiation.
• Other common findings:
  - Thrombocytopenia
  - Valvular heart disease
  - Livedo reticularis
  - Migraine
• Very few patients with primary APS go on to develop SLE
DIAGNOSIS

- Liquid phase coagulation assays (lupus anticoagulant)
  - APTT
  - Dilute Russell's viper venom time (DRVVT)
  - Kaolin clotting time (KCT)
  - Prothrombin time

- Solid phase ELISA assays (anti-cardiolipin antibodies).
TREATMENT

- Aspirin- inhibit platelet activation and/or warfarin as an anticoagulant.

- Low molecular wt Heparin

- The goal of the prophylactic treatment is to maintain the patient's INR between 2.0 - 3.0

- It is not usually done in patients who have not had any thrombotic symptoms.
• During pregnancy, low molecular weight heparin and low-dose aspirin are often prescribed.

• Women with recurrent miscarriage are often advised to take aspirin and to start low molecular weight heparin treatment after missing a menstrual cycle.

• In refractory cases, plasmapheresis can be of use.
LITERATURE REVIEW


Top of Basilar Artery Stroke with APS in 8 yr old

Department of Pediatrics, Government Medical College and Hospital, Postgraduate Institute of Medical Education and Research, Chandigarh India. 2001
THANK YOU