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Case Report

Diaphragmatic dysfunction: an unusual complication in HELLP syndrome

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ABSTRACT

Diaphragmatic dysfunction in HELLP [hemolysis (H), elevated liver enzymes (EL), low platelet count (LP)] Syndrome and Diaphragmatic dysfunction is an uncommonly reported association, wherein, patients who develop acute respiratory insufficiency poses a delay in weaning without any obvious cause. A high degree of clinical suspicion of sepsis induced diaphragmatic dysfunction needs to be kept in mind by the clinicians dealing with cases of HELLP Syndrome who may develop acute respiratory insufficiency without definite predisposing factors.

Keywords: Acute respiratory problems, Diaphragmatic dysfunction, HELLP syndrome

INTRODUCTION

Louis Weinstein in 1982 described a symptom complex called HELLP Syndrome [Hemolysis, Elevated Liver enzymes and Low Platelets] Acute respiratory insufficiency is an uncommonly occurring complication in HELLP syndrome, which may pose unusual delay in weaning from ventilator.[1,2] This complication is generally under-recognized, and diagnosis is frequently delayed or retrospective. Hence, there is a necessity for high degree of clinical suspicion in such cases with special emphasis on avoiding unduly hasty extubation.

CASE REPORT

A 22-year-old primigravida of 34(+) weeks amenorrhoea was admitted with feeling of breathless, dizziness and weakness. She had severe pallor, pedal oedema, heart rate (HR)-96/min, blood pressure (BP)-150/100 mmHg on Tab Labetolol 50mg twice daily. She had severe oligohydramnias and IUGR but insignificant past medical history. Her haemoglobin (Hb) was 3.0gm%(Microcytic Hypochromic Anaemia), PCV=9%, Total leukocyte count (TLC)= 22800/cmm (Polymorphs=90), Platelet Count=1.25Lac/cmm; Urine Albumin=+, Blood urea=78mg%, Serum creatinine=1.1mg%, Serum uric acid=4.5mg%, SGOT=96IU/L, SGPT=53IU/L and Alk phosphatase=321IU/L. She was transfused with 02 units fresh whole blood and planned for elective cesarean delivery.

Pre-anaesthesia assessment on next day revealed HR-86/min, BP-166/100mmHg, Respiratory rate(RR)-16/min, Hb=6.0gm%, PCV=17% TLC=15000/cmm(Polymorphs=89), Platelet Count=35000/cmm; Urine Albumin=++, Total Bilirubin=1.2mg%, SGOT=271IU/L, SGPT=132IU/L, Blood Urea=78mg%; Serum Creatinine=1.9mg% Serum uric acid=6.9mg%, Alk phosphatase=320IU/L and adequate urine output.

Around 11:30 hours on 3rd day, she suddenly developed generalised tonic-clonic seizures. She was breathing spontaneously but not responding to verbal commands.

HR was 56/minute, BP was 198/140mmHg, oxygen saturation (SpO2) on 100% O_2 by face mask (FM) was 86% and extremities were cold. She was resuscitated with I/V fluids, oxygen and taken up for emergency cesarean delivery under general anaesthesia using Inj Thiopentone (300mgI/V), Inj Succinylcholine (75mgI/V), Isoflurane and Inj Vecuronium (2mgI/V). A 2.0 Kg male baby was delivered. Since the baby had grunting, he was taken to NICU for further care. Uterus contracted well with Inj Carboprost Promethamine 250mcgI/M and 10IU Oxytocin infusion at 20drops/min. Post-operatively, muscle relaxation was reversed and extubation done. Though patient was fully awake, she failed to maintain adequate SpO2 on room air, hence put on supplemental oxygen through facemask. In ICU, she had HR=156/min and BP=148/98mmHg, Temperature=101OF, RR=36-37/min (paradoxical breathing pattern) and SpO2=70-80% on supplemental oxygen. Suspecting acute respiratory insufficiency, she was re-intubated and planned for brief elective mechanical ventilatory support along with Nitroglycerine (NTG) infusion @0.4mcg/kg/min, I/V antibiotics and continuation of fresh whole blood transfusion.

On 2nd post-operative day, her HR=134/min, BP=126mmHg, Temperature=102.2OF, RR=26-27/min (paradoxical breathing pattern), chest radiograph (PA view) revealed bilateral fluffy airspace opacities suggestive of Acute Respiratory Distress Syndrome (ARDS). Spontaneous but paradoxical breathing pattern (22-28/min) continued for next 48 hours. Repetitive weaning trials revealed a low tidal volume and low maximal inspiratory pressure. Her Hb=7gm%, TLC=16700/cmm (Polymorph=95), Platelet Count=75000/cmm, Urine Albumin=+, Total Bilirubin=1.2mg%, SGOT=200IU/L, SGPT=102IU/L, Blood Urea=82mg%, Serum Creatinine=1.1mg%. INR=1.6, PT=14sec (Control); 24sec (Test), PTTK=34sec (Control); 39sec (Test) and Toxic granules=Negative, hence Inj Vit K 1mgI/M also started.

Table 1: Causes of HELLP Syndrome.

S. No.	Disorders and associated complications
1.	Thrombotic disorders
a.	Thrombotic Thrombocytopenic Purpura (TTP)
b.	Hemolytic Uremic Syndrome (HUS)
c.	Sepsis and DIC
d.	Drug induced hemolytic anemia
2.	Consumptive disorders
a.	Acute fatty liver of pregnancy
b.	Sepsis and DIC
c.	Hemorrhage
3.	Others
a.	Connective tissue disorders
b.	SLE
c.	Anti Phospho Lipid Antibody Syndrome (APLA Syndrome)
d.	Pro-coagulant disorders

Patient responded well, and paradoxical breathing pattern gradually resolved by 4th post-operative day along with normalisation of coagulation profile and hemodynamic status. Mechanical ventilatory and NTG support was also weaned off on 5th post-operative day and discharged with a healthy baby on 10th post-operative day.

DISCUSSION

HELLP syndrome is a multi-systemic disorder with an incidence of 0.5 to 0.9% of all pregnancies.[3] Its etio-pathogenesis is not completely understood. It complicates pregnancy and has a poor prognosis. It was first described by Weinstein in 1982. Onset occurs in last trimester of pregnancy in 70% of pre-clamptic/eclamptic patients, and immediately after delivery in the rest.[4]

HELLP syndrome contributes to 6.7-70% perinatal mortality rate and 1-24% maternal mortality rate and increased rates of maternal morbidities such as pulmonary edema (8%), acute renal failure (3%), disseminated intravascular coagulopathy (DIC) (15%), abruptio placentae (9%), liver hemorrhage or failure (1%), ARDS, sepsis, and stroke (<1%).[4,5]

HELLP syndrome can present both as the primary expression of the preeclampsia process in pregnant patients or as a secondary phenomenon in patients with complicated sepsis, ARDS, renal failure, and multiple organ disease with DIC.[6] The differential diagnosis of HELLP syndrome is shown in Table-1. Sepsis is often associated with ARDS in the absence of infection. Detecting sepsis may be at times difficult as was evident in our patient also. Pre-eclampsia is known to be associated with HELLP syndrome and Acute Fatty liver of Pregnancy and sepsis is one of its dreaded complications. One of the organ whose function is severely compromised in sepsis is diaphragm.[7,8]

As investigated by Callahan and co-workers, sepsis induces a myopathy characterized by reduction in muscle force-generating capacity, atrophy and altered bioenergetics affecting both the respiratory muscles and the limb muscles. This leads to prolonged mechanical ventilation and difficult weaning.[9]

Several mechanisms have been reported to contribute to the development of diaphragmatic dysfunction in sepsis, including alteration in diaphragm mitochondrial function. Leigh and co-workers reported severe reduction in mitochondrial ATP generation capacity and an abnormally heightened production of free radical species by diaphragm. Both phenomena affect diaphragm force generation capacity thereby contributing to the prolonged need for mechanical ventilation in a previously asymptomatic HELLP syndrome patient susceptible to complicate into sepsis. This strengthens our belief of increased susceptibility of HELLP syndrome patients to complicate into sepsis (in the absence of documented

infection) to be a major contributory cause in developing diaphragmatic dysfunction.[10]

CONCLUSION

Since, HELLP syndrome may be a secondary phenomenon of a co-existing incipient sepsis which may be difficult to detect at times. A high degree of clinical suspicion of sepsis induced diaphragmatic dysfunction thus needs to be raised in HELLP Syndrome patients developing acute respiratory insufficiency without definite predisposing factors. An early clinical suspicion of nascent/incipient sepsis without frank evidence of infection, importance of avoiding undue haste in extubation and continuing elective mechanical ventilation are emphasized in patients of HELLP syndrome complicating into acute respiratory insufficiency and diaphragmatic dysfunction.

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