e-ISSN: 0975-5160, p-ISSN: 2820-2651

Available online on www.ijtpr.com

International Journal of Toxicological and Pharmacological Research 2022; 12(10); 130-132

Original Research Article

Anaesthetic Management of Gaucher's Disease in Obstetric Patient

Das Arunima[1], Khatun Ayesha[2], Sarkar Debananda[3], Sarkar Manabendra[4]

[1]PGT, Department of Anaesthesiology, NRS Medical College & Hospital, Kolkata
[2]Senior Resident, Department of Anaesthesiology, NRS Medical College & Hospital, Kolkata
[3]Assistant Professor, Department of Anaesthesiology, NRS Medical College & Hospital, Kolkata
[4]Professor, Department of Anaesthesiology, NRS Medical College & Hospital, Kolkata

Received: 30-08-2022 / Revised: 28-09-2022 / Accepted: 04-10-2022
Corresponding author: Dr. Arunima Das
Conflict of interest: Nil

Abstract

As we know Gaucher is the most common form of lysosomal storage disorder due to deficiency of the beta glucocerebrosidase enzyme. This condition may be a challenge for both anaesthesiologist and obstetrician as there is abnormal coagulation profile and multiorgan involvement in obstetric condition. Our aim was to report divergent management of "Gaucher disease" depending on patient's characteristics.

Keywords: Gaucher's disease, General anaesthesia, Pregnancy

Introduction

Pathology of Gaucher disease results from the accumulation of glucocerebrosides due to deficiency of glucocerebrosidase enzyme [1] in the reticuloendothelial system. This disease has three variants. Out of these three type I i.e adult, chronic, non neuropathic is the most common form. No neurological involvement is seen in type I like in type II and III Gaucher's disease.

Mutation is 1226G (N3705) [2]. Patient often come with hepatosplenomegaly and combination of "anaemia, leucopenia and thrombocytopenia". Type I Gaucher disease may put an anaesthesiologist to face difficult situations as there is coagulation disorder and multiple organ involvement in obstetric condition.

These factors have an effect on mode of delivery and also on consequences of anaesthetic implementations during delivery. Our target was to handle the situation according to the patient's condition and to report the management procedure.

Case

A 27 year old primi gravida (Height 148 cm, weight 44 kg) known case of Type I Gaucher disease got admitted in our institution at 36 weeks of gestation with IUGR, foetal distress and less liquor. She had past history of walking inability due to bodyache, abdominal protruberence and her height was not appropriate according to her peers group during her childhood period. That time she

was diagnosed with the Gauchers disease after proper investigations.

All of her symptoms decreased when she attended menarche (at 12 years of age). There was no history of Gaucher disease in her family. Beside this she had hypothyroidism but on irregular medication (LT4 75mg). No history of diabetes mellitus, hypertension, shortness of breath, syncopal attack, seizure disorder. On pre anaesthetic check up no cardiorespiratory abnormality was present but on palpation the hepatomegaly (4 fingers below costal margin) and the splenomegaly (up to umbilicus) found. No spinal abnormality detected. After her conception, she regularly visited Antenatal care unit and she was advised to visit haematology clinic of our institute with her bone marrow assay report. She was treated with Cerezyme 60 IU/kg i.v once in two weeks as a part of enzyme replacement therapy during her pregnancy period [3,4]. The blood investigation report showed that she was chronic anaemic and chronic thrombocytopenic. Her haemoglobin was 8.1 gram/dl and platelet was 40,000/mm3 on the day of operation. No history of bleeding episode. Fibroscan showed the early cirrhotic changes in liver. Electrocardiogram and Echo 2D was within normal limit (LVEF 65%). Both liver and renal function test was within normal limit [5,6].

We considered this case under general anaesthesia [7] because of thrombocytopenia there was risk of hematoma formation on the site of Regional anaesthesia.

As there was a chance of organ injury due to the organomegaly [8] we transfused 1 unit PRBC, 4 units FFP pre operatively as a precautionary measurement. The patient was kept in supine position with left lateral uterine displacement to minimize the aortocaval compression in addition to routine monitors. (NIBP, 5 leads ECG, Pulse oxymetry).

Pre oxygenation was done with 100% oxygen for 5 minutes. Premedications given as per body weight of the patient (glycopyrrolate+ ondansetron). Modified rapid sequence intubation was performed after giving induction agent (propofol) and muscle relaxants (succinyl choline). Patient was intubated by 6.5 mm ID cuffed endotracheal tube under direct laryngoscopy in a gentle manner within 10 seconds in a single attempt. After confirming the correct position of tube, it was fixed at angle of mouth. Maintainance was done with nitrous oxide and oxygen in 1:1 ratio. Inhalation with Sevoflurane (MAC 1) was given. Injection Atracurium was given intermittently for muscle relaxation. Paracetamol 1 gram was given before incision. Incision delivery time was five minutes. We gave oxytocin (10U) i.m and 10 U with 500 ml Ringer lactate and injection fentanyl 100 microgram i.v. In intraoperative period systolic BP was in a range of 120-136 mmHg and diastolic BP was in a range of 70-86 mmHg. Heart rate was within 80-100/minute. Injection Methargin 1 ampule was given intramuscularly after delivery of baby as the uterus was flabby. Intravenous fluid ringer lactate 1500 ml+ 1U PRBC given in intraoperative period. The operation was lasted for 1 hour. Blood loss was approx. 750-800 ml.

Bilateral TAP block was given for postoperative analgesia 10 ml on each side. Residual paralysis was reversed by giving reversal agent (Neostigmine + Glycopyrrolate). Tracheal extubation done when neuromuscular function reappear. Post operatively patient was doing well. She was shifted to HDU for the better care and careful observation done regarding vitals and new onset postpartum haemorrhage [9]. She was given oxygen support via facemask with 6 lit/minute for a four hours of duration.

Platelet concentrate 4U and FFP 4U was transfused within 2 hours of LUCS. Another 1U of PRBC was transfused on next day.

Proper analgesic management was given through PCA. After 24 hours of careful observation she was shifted to post natal ward. After 7 days of LUCS she was discharged along with the healthy female baby weighted 2.1 kg.

Discussion

Gauchers disease is a rare disease, specially in pregnant woman so it creates challenges for anaesthesiologists for choices of anaesthesia between general and regional. Any of them can be used for management of this case. Gauchers disease is a multiorgan disease,so through pre operative check up should be carried out to find any organ (lung, liver, spleen, bonemarrow) involvement and specially whether central nervous system is involved or not. Hepatic involvement (Hepatomegaly) is the most common sign of 3 types of Gauchers disease and this may lead to cirrhosis of liver which further progresses to hepatic fibrosis (Fibroscan of liver). Splenomegaly can cause hypersplenism which can produce thrombocytopenia, anaemia. So, haematological status assessment in pre operative period is of primary concern.

Thrombocytopenia may be sufficiently severe to preclude the regional anaesthesia. Coagulation factor deficiencies are common in this population and therefore coagulation profile, bleeding time, clotting time should be carried out. Increase in bleeding time due to alteration of the clotting parameters is result of platelet function abnormality. Our patient has no coagulation abnormality. Anaesthetic management of parturient who is having Gauchers disease basically depends on the clinical manifestations and patient's physical condition.

Outcome

With this management patient recovered well without any complication and discharged with healthy baby.

Referrence

1. Brady. R .O., Kanfer, J.N. & Shapiro, D. (1965) Metabolism of glucocerebrosidase. II: Evidence of an enzymatic deficiency in Gaucher's disease. 1965 Jan 18; 18:221-5.
2. Beutler E, Gelbert T. Gaucher disease mutations in non Jewish patients. Br J Haematol. 1993; 85: 401-5
3. Hollander C E, Levi M, Bernard's F, Aerts J M, van Oers M H. Coagulation abnormalities in type 1 Gaucher disease are due to low-grade activation and can be partly restored by enzyme supplementation therapy. Br J Haematol. 1997; 96: 470-476
4. Barton N W, Brady R O, Dambrosia J M, *et al*. Replacement therapy for inherited enzyme deficiency: macrophage-targeted glucocerebrosidase for Gaucher's disease. 1991 May 23; 324(21):1464-70.
5. Epstein Y, Eisenberg V, Granovsky-Grisaru S, *et al*. Pregnancies in Gaucher disease: A 5-year study. Am J Obstet Gynecol. 2004; 190: 435-441.
6. Goldblatt J. Beighton P. Obstetrics aspect of Gaucher's disease. Br J Obstet Gynaecol. 1985; 92: 145
7. Ioscovich A, Elstein Y, Halpern S, Vatashsky E, Grisaru-Granovsky S, Elstein D. Anaesthesia for obstetric patients with Gaucher disease: survey and review. Int J Obstet Anesth. 2004; 13: 244-50.
8. Jane's S.P. Stromeyer F.W. Chang C. Barratrger J. A Liver abnormalities in patients with Gaucher's disease. Gastroenterology. 1981; 80: 126
9. Grabowski G. Recent clinical progress in Gaucher disease. Curr Opin Pediatr. 2005; 17: 519-24.