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Anesthetic Management of a Rare Congenital Limb Anomaly with Rh-Negative Pregnancy: A Cesarean Section Case Report

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Abstract: ***We present a rare and challenging case of a pregnant woman with tetraphocomelia, an extremely rare congenital limb anomaly characterized by the absence or severe hypoplasia of all four limbs, scheduled for elective cesarean section. The patient was Rh-negative and had a history of previous lower segment cesarean section (LSCS). This unique combination of anatomical deformity, immunohematologic considerations, and obstetric history required a highly individualized multidisciplinary approach to perioperative anesthetic planning. Key considerations included positioning, vascular access, neuraxial anesthesia feasibility, airway management, and intraoperative monitoring. Phocomelia is a rare birth defect that results in significant abnormalities in limb development. It often involves underdeveloped or missing upper limb bones, and the fingers may be fused together. In the most severe cases, known as tetraphocomelia, both the arms and legs are absent, making the hands and feet appear to be directly connected to the body. This condition can be caused by genetic factors or exposure to certain drugs. Genetically, it is usually inherited as an autosomal recessive trait and is linked to various chromosomal abnormalities. It may also arise from spontaneous genetic mutations.***

Keywords: tetraphocomelia, congenital limb deformity, cesarean anesthesia, multidisciplinary management, genetic mutation

1. Case Report

A 32 year old case of Tetraphocomelia with $G_3P_1L_1A_1$ with 37^{+5} weeks period of gestation with Rh negative pregnancy with previous history of LSCS with oblique lie and scar tenderness posted for caesarean section. Brief pre anaesthetic examination was done. Patient gives history of previous LSCS 2 ½ years ago, which was under Sub arachnoid block with i. v sedation and was uneventful. H/O 3 pints PRBC transfusion 5 years ago i/v/o low Hb of 4g/dl. No reactions noted during transfusion. No comorbidities noted in the patient. General physical examination showed no presence of pallor, icterus, clubbing, cyanosis, lymphadenopathy or edema. Breast, Thyroid and Spine was found to be normal. B/L upper and lower limb deformity noted. Vitals: PR: 98bpm; BP-110/70mm Hg Investigations (17/11/21): Hb-11.4g/dl; TC-8900 cells/cumm; Platelets: 1.75 lakhs/cumm; Blood group-'O' negative Coagulation profile: PT-13.3; APTT-41.2; INR-1.01

Systemic examination:

RS: B/L NVBS heard, no added sounds

CVS: S1 S2 heard, no murmurs

P/A-Gravid uterus corresponding to the size of the uterus.

CNS-HMF found to be normal with no focal neurological deficits.

Case was accepted under ASA-II

Obstetric History

A1-Spontaneous abortion at 1 ½ months. No suction evacuation done

P1-A healthy female baby delivered by LSCS, 2 ½ years ago, BW-3.4kg. Inj Anti-D 300mcg was given after the delivery of the baby

Present pregnancy: Patient was a booked case with regular ANC visits. Antenatal history-Uneventful

Anomaly scan was found to be normal

Lab investigations were within normal range

Intraoperative anaesthetic management consisted of non-invasive blood pressure monitoring via a standard BP cuff on left upper extremity. Pulse oximeter was connected to patient's right finger and ECG leads were connected. Due to the patient's history of difficult venous access and possibility of abnormal upper extremity vascular anatomy, CVP line was secured into the Right Internal jugular vein under sterile aseptic precautions with 12F size Triple lumen central venous catheter by means of Modified Seldinger's technique under USG guidance.

The intraoperative Blood Pressure, Heart rate, Oxygen Saturation, Respiratory Rate and End Tidal Co2 were monitored and recorded.

Patient was placed in the Left lateral position. Under sterile aseptic precautions, Spinal anaesthesia was given at L3-L4 IVDS by using 25-G Quincke's needle with 2ml of 0.5 % Bupivacaine (H).

A single live healthy male baby of weight 2.92 kg was delivered. Baby cried immediately after birth. With Apgar scores of 9 (1 min) and 10 (5 min).

Inj. Oxytocin 15U slow iv was given to aid uterine contraction. Blood pressure and normothermia was maintained throughout the procedure. Patient was haemodynamically stable. Blood loss was approximately 450 ml. A total of 1200 ml of Ringer Lactate solution was given

in the perioperative period. After the end of the procedure, patient was shifted to the post operative care ward with special care and assistance.

2. Discussion

Phocomelia can either be genetically inherited or drug-induced. In its genetic form it is transmitted as an autosomal recessive trait which is linked to several chromosomal mutations. Spontaneous gene mutations have also been recognised. Drug-induced cases of phocomelia syndrome can be mostly associated with maternal ingestion of the drug thalidomide by expectant females (Thalidomide syndrome). Thalidomide was developed in Germany in 1954 and gained widespread use as treatment for morning sickness in early pregnancy. It was also used as a sedative and cough suppressant. It became apparent in the 1960's that thalidomide treatment resulted in severe birth defects in thousands of children.

Other commonest anomalies associated with phocomelia include thoracic scoliosis, upper thoracic hemivertebrae, and cardiomegaly.

Differential diagnosis of phocomelia are Sporadic phocomelia, Holt-Oram syndrome, Thrombocytopenia-absent radius syndrome (TAR syndrome), Roberts syndrome, Thalidomide-induced phocomelia.

Sporadic phocomelia is a very rare genetic disorder inherited as an autosomal recessive trait or as a result of spontaneous mutation. Thrombocytopenia-absent radius syndrome (TAR) is characterized by a low platelet count, an absent radius, a hypoplastic thumb and cardiac abnormalities.

Holt-Oram syndrome may be seen both as an autosomal dominant disorder and as a result of spontaneous genetic mutation. This syndrome is characterized by abnormal limb development that affects mostly the forearm and the carpal bones of the wrists. Characteristic features include a hypoplastic thumb or a thumb that looks like a finger. Frequently, the radius is missing and the humerus is underdeveloped. The clavicle and scapula may be affected. Three-quarters of the patients with Holt–Oram syndrome have cardiac problems that may include atrial septal defects or ventricular septal defects.

Anesthetic Challenges in this Case:

- **Monitoring of Blood Pressure:** Non-invasive blood pressure monitoring may be difficult due to limb deformities or anatomical limitations.
- **Venous Access:** Securing intravenous access may be challenging owing to abnormal limb development and limited venous sites.
- **Anticipated Difficult Airway:** Airway management is expected to be difficult, necessitating careful attention to patient positioning due to restricted mobility. Special precautions must be taken during handling and positioning to ensure both patient safety and optimal access.
- **Cardiovascular Monitoring:** Invasive (e. g., transesophageal echocardiography) or non-invasive (e. g., IVC diameter measurement) monitoring may be required to guide fluid management during the perioperative period.
- **Choice of Anaesthesia:** General anaesthesia is generally preferred due to the frequent association of phocomelia with other congenital anomalies. However, there are no absolute contraindications to regional anaesthesia if appropriate.

3. Summary

A 32-year-old gravida with tetraphocomelia, Rh-negative status, and previous LSCS was scheduled for elective cesarean section at 37+5 weeks of gestation.

She had an uneventful antenatal course, with a history of prior spinal anaesthesia and blood transfusion without complications.

Due to anticipated difficulty in venous access, a central line was secured under ultrasound guidance. Spinal anaesthesia was administered successfully, resulting in the delivery of a healthy male baby. Key anaesthetic challenges included difficult venous access, airway management, and positioning, all managed effectively with a stable intraoperative course.

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