

CASE REPORT

Cesarean Section in a Female with Turner's Syndrome: A Case Report

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ABSTRACT

Turner syndrome, a chromosomal disorder characterized by partial or complete loss of X chromosome, presents unique challenges during pregnancy and anesthesia management due to associated comorbidities such as cardiac anomalies and musculoskeletal abnormalities. A 30-year-old primigravid woman with mosaic Turner syndrome underwent an emergency cesarean section at 38 weeks and 2 days of gestation due to arrest at the second stage of labor with fetal distress. The pregnancy was considered high-risk due to gestational hypertension, hypothyroidism, and premature rupture of membranes (PROM). Physical characteristics of Turner syndrome, such as short stature, low-set ears, and a broad chest, were present, though no major cardiac abnormalities were identified. Spinal anesthesia was administered using 2 ml of 0.5% bupivacaine, achieving a T5 sensory block. The procedure was uneventful, resulting in the delivery of a healthy male infant. Postoperative recovery was smooth, with no cardiac complications. This case emphasizes the importance of thorough preoperative evaluation, vigilant monitoring, and tailored anesthetic techniques to minimize risks.

KEYWORDS

• Mosaic turner syndrome • Ceasarian section • Chromosomal abnormalities

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INTRODUCTION

Turner's syndrome is a genetic disorder caused due to a missing or a partial 'X' chromosome. Apart from stunted growth and multiple physical characteristics, girls born with Turner's syndrome exhibit an incomplete sexual development. Women with Turner's usually have absent or delayed pubertal development, accelerated degeneration of oocytes possibly leading to infertility.^[1] On the contrary, mosaic Turner syndrome occurs due to a faulty cell division during early fetal development resulting in a varied proportion of cells with 'XO' and 'XX' genotypes.^[2] Although rare, women suffering from Turner mosaic have been reported to have conceived naturally, though with a higher incidence of still birth, miscarriages, birth defects in the baby and multiple maternofetal complications during pregnancy.^[3]

CASE REPORT

We present a case of a 30-year-old female, primigravida, with history of primary infertility posted for an emergency Caesarean section in view of arrest in 2nd stage of labour with a decelerating fetal heart rate. Patient had an average built with a body mass index (BMI) of 23.78 kg/m² (height 145 cm, weight 55 kg). On examination, the patient had low-set ears, scanty and short hair growth on the scalp since birth, a low hairline, short neck, and a broad chest [Fig. 1]. She was diagnosed with hypothyroidism 1 month ago, and was taking 25mcg thyroxine per day since then. On rupture of membranes, she approached a nearby district hospital, from where she was referred to our institute in view of precious pregnancy with suspected mosaic Turner syndrome. During the pre-anaesthetic evaluation, her pulse rate was 80 beats per minute, blood pressure 130/80 mmHg, and oxygen saturation (SpO₂) 99%. Head and neck examination revealed normal, intact dentition, a Mallampati III upper airway [Fig. 2], and a normal range of neck motion. Thyromental distance was 6.8 cm, upper lip bite test was class 2. On examination, she had mild pallor. Systemic examination revealed no clinical abnormality in respiratory, cardiac or central nervous system. The electrocardiogram showed normal sinus rhythm. No record of ANC visits was found. Bleeding and clotting times were normal, and her urine

tested negative for albumin. Screening for gestational diabetes was negative. No other investigations were available.



Fig. 1

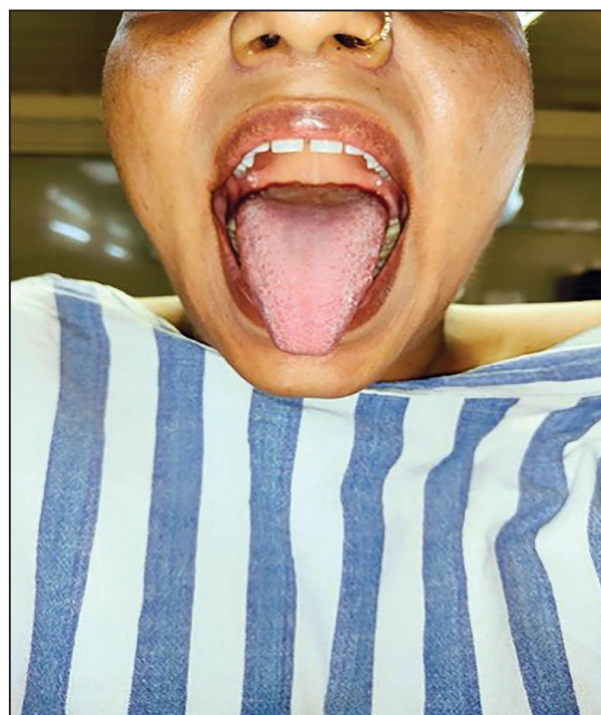


Fig. 2

In view of foetal heart rate decelerations, it was decided to proceed with the Caesarean section. The operation theatre was prepared according to difficult airway algorithms. The crash cart was equipped with a charged defibrillator, inotropic agents, vasopressors, antihypertensives and resuscitation drugs. After confirming NPO status, she was taken to the operating room. Standard ASA monitoring was attached, and an intravenous line was secured. Under aseptic conditions, the subarachnoid space was accessed, and 2 ml of 0.5% bupivacaine (heavy) was given intrathecally via a 25-gauge Quincke's spinal needle. A T5 sensory block to pinprick was achieved after five minutes.

All cardiovascular parameters remained stable during surgery. A healthy male child, weighing 3000 g and with normal Apgar scores at 1 and 5 minutes, was delivered. Adequate uterine contraction was achieved with an oxytocin infusion after the delivery of the placenta. Blood loss was approximately 500 mL, and urine output was 200 ml. After recovery room criteria were met, the patient was transferred to the ward. Postoperative analgesia was provided with TAP block and intravenous Paracetamol. Postoperative echocardiography was performed to detect any cardiac abnormalities, but the results showed none, with normal valvular and ventricular function and an estimated ejection fraction above 60%. She recovered well and was discharged home three days after the operation.

DISCUSSION

Turner syndrome (TS), first described in 1938, is the most common chromosomal abnormality in females, with an incidence of approximately 1 in 2500 to 1 in 2500 live female births. The condition arises due to anomalies in the sex chromosomes, with the classic form involving complete monosomy of the X chromosome (45, X), and the mosaic form involving two or more cell lines with different karyotypes, such as 45, X/46, XX, 45, X/47, XXX, or 45, X/46, XY. Mosaicism is detected in a significant proportion of TS cases and is often associated with milder symptoms.^[5] A standard 20 - cell karyotype is required to diagnose Turner syndrome. We were unable to perform karyotype study due to financial constraints. Hence the diagnosis was made based on clinical features and

managed accordingly.

Women with Turner syndrome pose a multitude of challenges for the attending anesthesiologists due to their specific anatomical and physiological changes. Establishing an artificial airway tends to be difficult owing to short and wide neck, limited neck movement, limited mouth opening, high arched palate and mandibular hypoplasia. These patients may present with cardiorespiratory (coarctation of the aorta, bicuspid aortic valve, aortic stenosis, hypertension), musculoskeletal (scoliosis), Renal (Horseshoe kidney or other structural defects) and gastrointestinal (gastroesophageal reflux) anomalies. Other endocrine and metabolic abnormalities like hypothyroidism, obesity, diabetes mellitus and dyslipidemia may frequently be co-existent in such patients.^[6,7] Most women with TS experience infertility due to gonadal dysgenesis, although about 10% achieve spontaneous menarche, and a small proportion (5-6%) experience spontaneous pregnancy, particularly those with mosaic karyotypes.^[5]

Although Turner syndrome is the most common chromosomal anomaly in females, these patients presenting for surgery is quite rare. Decision regarding mode of anesthesia depends on the type of surgery, physical features, associated cardiac, respiratory and renal anomalies. Any elective surgery demands a thorough pre-anesthetic clinical evaluation, complete hemogram, chest X-ray, ECG, 2-D echo, renal function tests and liver function tests.^[8] We were confronted with an emergency situation wherein mode of anesthesia was chosen weighing the risks and benefits based on available resources and expertise. In our patient, we chose spinal anaesthesia as clinical examination did not reveal any abnormality. A written informed high - risk consent was taken for anesthesia and surgery. A difficult intubation cart and cardiac resuscitation tray were kept ready.

Turner syndrome is often associated with spinal issues like kyphoscoliosis, increased lumbar curvature, and short stature, which can make it harder to perform spinal or epidural anaesthesia. These issues may increase the risk of complications such as accidental punctures or uneven drug distribution^[9]. Michala L *et al.* reported using a low-dose combined spinal-epidural (CSE) technique in a patient with mosaic Turner syndrome.^[10,11] Regional

anaesthesia has several benefits: it avoids the challenges of a potentially difficult airway, reduces the risk of vascular complications during pregnancy, and limits the stress on the heart and blood vessels that can occur with intubation and anaesthesia recovery. The excellent analgesia promotes early recovery and breast feeding.^[12]

There can be no simple algorithm governing anaesthetic management in a patient with Turner syndrome. Every case requires a personalized management protocol. Although the surgery went uneventful in our case, the anaesthesia team needs to be prepared for life threatening complications like aortic dissection. A thorough knowledge of clinical features, pathophysiology and possible complications is mandatory for successful management.

CONCLUSION

In summary, this report highlights the unique anaesthetic and medical considerations for patients with Turner syndrome. Women with mosaic Turner syndrome who can conceive naturally should have regular heart check-ups and blood tests, as their pregnancies are high-risk. With careful management, cesarean section for these pregnancies can be successfully handled using low-dose sequential combined spinal-epidural anesthesia.

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