Journal of Cardiovascular Disease Research

ISSN: 0975-3583, 0976-2833 VOL15, ISSUE 1, 2024

Anaesthetic Management of Patient with Marfan Syndrome during Caesarean Section: A Case Report

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Abstract

Marfan syndrome (MFS) is an autosomal dominant connective tissue disorder mainly involving the cardiovascular, skeletal and ocular systems. Cardiovascular manifestations, such as aortic dilatation and dissection, are responsible for 90% of deaths attributed to MFS. Management of anesthesia is challenging in pregnant patients with Marfan syndrome and no ideal technique has been defined.

KEYWORDS- Marfan syndrome, Pregnancy, Anaesthetic management

INTRODUCTION:

On examination, she had mild pallor and arachnodactyly (Fig 1) with arm Marfan syndrome is primarily a connective tissue disorder due to mutation of the gene encoding fibrillin-1(FBN1). It affects males and females equally and its incidence is 1 to 2 per 10,000. Fibrillin is a matrix glycoprotein essential for the formation of microfibrils that regulate the formation and repair of connective tissue. It is characterized by dilatation of the aortic root, dislocation of the ocular lens, decreased skeletal muscle mass, mitral valve prolapse, long bones, and other skeletal abnormalities. [1 ,2]

Microfibril abnormalities in the myocardium may affect normal atrial to ventricular conduction. Pregnancy aggravates the condition due to increased cardiac output. Cardiac output reaches a maximum during labor (release of catecholamines in response to pain) and immediately after delivery due to which during intrapartum and postpartum period patients with Marfan syndrome are at increased risk of life-threatening aortic dissection, especially when the aortic root diameter is larger than 40 mm.[3-6].

Management of anesthesia and analgesia is challenging in patients with Marfan syndrome and

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no ideal technique has been defined. In healthy women, neuraxial techniques are the techniques of choice for analgesia during labor or for anesthesia during cesarean delivery. However, lumbar spine deformities or a history of lumbar spine arthrodesis are common in patients with Marfan syndrome (MFS), which may preclude successful epidural catheter or spinal needle placement. Similarly, the presence of dural ectasia is associated with an increased risk of dural puncture during epidural catheter placement for labor analgesia or ineffective spinal anesthesia during cesarean delivery.[7,8] We report a case of 27 year old primigravida with marfan syndrome who required anaesthetic care.

Case Report:

A 27 years old, primigravida, referred from peripheral hospital, 175 cm tall and weighing 55kg was posted for elective caesarean section (CS) at 40 weeks of gestation. She did not give any previous history of medical illness related to cardiovascular system. During her second trimester of pregnancy, she gave history of occasional palpitation with mild exertional dyspnoea (NYHA II) which was gradually progressive with increasing trimester. Her family history suggested that her mother had similar complaints and died at the time of her vaginal delivery at home.

On examination, she had mild pallor and arachnodactyly (Fig 1) with arm span more than her height (arm span 177cm, height 175cm) and mild kyphoscoliosis. Her MET(metabolic equivalents of task) Score was 4. Her airway examination showed high arched palate (Fig 2), buck teeth (Fig 3) with Mallampati class II. On cardiovascular examination, we found wide pulse pressure on palpation of radial artery, an early diastolic murmur on auscultation of precordium. ECG had a sinus rhythm with a heart rate of 130 beats per minute. The patient was normotensive(BP 120/84mmHg). Patient was evaluated by cardiologist and echocardiography showed aortic root dilatation (root diameter 50 mm) with mild aortic regurgitation (AR), mild mitral regurgitation(MR), mild TR, bicuspid Aortic valve and grade II mitral valve prolapse (MVP). Left ventricular ejection fraction was 45-50%.Cardiothoracic surgery opinion was also taken .Tablet metoprolol 50 mg twice a day was started for 3 days prior to surgery by the cardiologist. Her hemoglobin was 8.8 g%, prothrombin time 12.4 (INR 0.92) and thyroid function tests and serum electrolytes values were within normal limit. She was planned for elective CS. The patient and her relatives were informed about the anaesthetic technique and written informed high risk consent was obtained. In the morning of surgery, patient was administered dose of tablet metoprolol, tablet pantoprazole 40 mg in night and in morning before surgery. Adequate blood and blood products were arranged. A multidisciplinary

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involvement of anesthesiologist, gynaecologist and cardiac surgeon were requested in anticipation of requirement for cardiopulmonary bypass. We planned for combined spinal and epidural anaesthesia. With monitoring of 5 lead electrocardiogram (ECG), noninvasive blood pressure and pulse oximetry, baseline recorded HR 126/min, BP 126/80 and Spo2 100%. Two intravenous line secured with 18 G cannula and IV □uid (ringers lactate) was started. Preloading was done with 250 ml of IV □uid over 40 minutes prior to anaesthesia. In sitting position epidural anaesthesia was performed by midline approach with the Tuohy's needle size 18 G and epidural catheter size 20 G was inserted in L2-L3 intervertebral space. Subarachnoid block was achieved through Quincke spinal needle size 23 G at L3-L4 intervertebral space with 1.8ml of bupivacaine heavy 0.5%. Patient was made supine, wedge was applied under the right buttock. Adequate anaesthesia level upto T10 was achieved within 4 minutes. Caesarean section was performed, the healthy male newborn (2.5 kg) with immediate cry was delivered with APGARS score in the first minute was 9, and it was 10 after five minutes. There was single episode of hypotension (BP-72/52 mmHg) then 20 microgram phenylepinehrine was given which was sufficient to normalize the blood pressure. There was no evidence of intraoperative and postoperative complications. Total duration of anaesthesia was 90 minutes, total fluid administered was 1.5 litres and one unit of blood was administered intraoperatively after delivery of baby. The epidural catheter was removed 24 hours after caesarean section. First dose of epidural drug was given after 45 min when patient complained of pain with 8ml of 0.25% ropivacine and second dose 0.125% after 6 hours when VAS score was 5. The patient was kept under observation for 3 hours in the PACU and then transferred to ward. The postoperative period was uneventful .One month after the surgery patient was posted for Bentall's procedure (ascending aorta repair) in CTVS OT. Intraoperative and postoperative period was uneventful. She was extubated 24 hours after surgery and discharged after one week of surgery.

DISCUSSION:

Marfan syndrome is named after Antonie Marfan , a French Paediatrician who first described Arachnodactyly in 1896 in the bulletin of medical society of paris in a 5 year old girl . MFS is an autosomal dominant disorder of the connective tissue related to mutation of the gene for fibrillin, a glycoprotein that is the major component of extracellular microfibrils,

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Fig 1 Long Upper limb digits

Fig 2 High arched palate

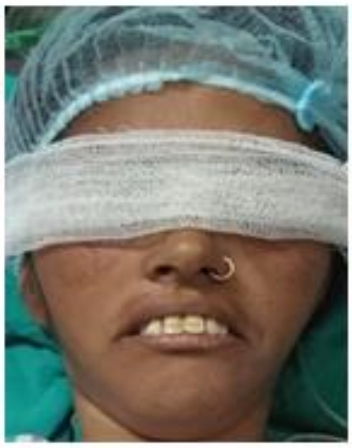

Fig 3 Crowding of teeth

Fig 4 Upper to lower segment ratio < 0.86

Fig 5 Arm span and height ratio 1.01

(chromosome 15). MFS involves different organs and systems with varying severity: for this reason, its diagnosis is mainly clinical, instrumental and molecular. It is based on the observation of the revised ghent criteria. [9] Thorough preoperative evaluation should be done with identification of benefits, adverse effects and implications related to any intervention. Cardiovascular system is most commonly involved in patients with marfan syndrome. The most common manifestation of cardiovascular system is Aortic root dilatation In addition to the cardiac abnormalities, dificult airway due to high arched palate, kyphoscoliosis (reduction in total lung capacity) and high chances of temporomandibular joint dislocation are the main concerns of anaesthesiologist.

As in our case, patient was 40 weeks primigravada so uctuation in hemodynamic parameters

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secondary to pain and anxiety of labor may have negative effects on the cardiovascular system; high blood pressure tends to develop aortic aneurysms due to weakened vascular media in patients with MFS, and myocardial ischemia and heart failure can also be caused by an increased myocardial oxygen demand resulting in high blood pressure; thus, the main goal is to prevent high blood pressure.[10] Drastic reductions in preload should be avoided. Spontaneous labor and vaginal delivery were recommended when aortic root diameter was less than 40 mm. Since our patient had aortic rot diameter more than 40mm and the spontaneous birth determines increase in blood pressure during contractions, due to which, caesarean section was planned.

General anaesthesia causes blood pressure variations during intubation; therefore regional anaesthesia seems preferable for our patient because of slow onset and gradual progression of epidural block. Combined spinal-epidural anaesthesia is preferred over general anaesthesia for caesarean section in patients with MFS because combined spinal-epidural anaesthesia provides excellent hemodynamic stability, and adequate postoperative pain control may be obtained via epidural analgesia. However, many cases of spinal anaesthesia failure have been reported in Marfan patients, possibly due to dural ectasia.[11,12] Few cases of incorrect or inadequate spread of intrathecal local anaesthetic in patients with this syndrome have been described. Lacassie et al.[13] performed continuous spinal anaesthesia in two patients with an incrementally increased dose of bupivacaine, but they stopped further administration of bupivacaine after 21ml for the fear of potential neurological damage. They also reported an irregular distribution of spinal anaesthesia due to unpredictable and inadequate spread of intrathecal local anaesthetics in patients with MFS. One of the most important factors in□uencing the height of the block in patients receiving spinal anaesthesia is the volume of CSF in the lumbosacral space, which contributes to the variability in the spread of spinal block. Kimet al.[14] reported a successful perioperative management of a patient with MFS and dural ectasia for caesarean section using epidural anaesthesia, adequate level of anaesthesia was achieved 30 min after the epidural injection of 27ml of 2% lidocaine with epinephrine (1 : 200) and fentanyl (100mcg).

Management of aortic root dilatation involves medical therapy as well as surgery. Beta-blockers reduces the heart rate, blood pressure, the risk of aortic dilatation and cardiac complications, but they seem to increase the tone and uterine contractility, and they might reduce the □ow in the umbilical artery causing low birth weight infants.

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CONCLUSION

The evaluation of pregnant women with MFS requires multidisciplinary management with a close cooperation between gynecologist, cardiologist, anesthesiologist, and neonatologist. Pregnancy should be programmed after complete evaluation of the patient and the definition of specific risks. Relevant is the echocardiographic assessment of aortic root dilation. During pregnancy, the obstetric management is not significantly different, but it is burdened with a higher frequency of premature rupture of membranes, the side effects of the drugs used, where indicated, for the prevention of aortic rupture, and the risk of aortic dissection. Regional anaesthesia has been successfully used during caesarean section, but there is a significant probability of erratic and inadequate intrathecal spread of local anaesthetics, most likely as a result of dural ectasia. In these patients, epidural anaesthesia may be a particularly useful technique during caesarean delivery because it allows adequate spread and action of local anaesthetic and controlled onset of anaesthesia, analgesia, and sympathetic block with low risk of complications. We report the perioperative management of a patient with MFS who underwent successful caesarean delivery using epidural technique anaesthesia.

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