Sequence of events which led to significant hypoxemia in a patient with Marfanoid features undergoing Elective Cesarean Section under general anaesthesia: A case report

Dr. V. Devadas; Dr. H. Dharmakeerthi

Registrar in Anesthesia[1]; Consultant Anesthetist[2], Colombo North Teaching Hospital Sri Lanka

Abstract:

The airway and respiratory system abnormalities seen in Marfan syndrome pose a significant challenge to the anaesthetist. Here, we discuss the chain of events that occurred during the induction of a primi mother, with Marfanoid features complicated with low platelet count and anaemia due to bone marrow hypoplasia. We then discuss the concerns of Marfan Syndrome and its features concerning anaesthetic management.

KEYWORDS: Marfanoid features; Scoliosis; difficult airway; Marfan Syndrome; Pneumothorax; restrictive type lung diseases

INTRODUCTION

The important abnormalities from an anesthetic standpoint in Marfan's syndrome or Marfanoid features include a high arched palate (airway implication), pectus excavatum, hyperextensible joints (possible positioning implications), and kyphoscoliosis (difficulties with neuraxial anesthesia and restrictive lung disease).

*Correspondence: Dr Vimalya Devadas

Email : vdevadas23@gmail.com

https://orcid.org/0009-0008-7328-6122

Received: 07/03/2023

Accepted: 30/09/2023

DOI: https://doi.org/10.4038/slja.v32i1.9189

The anesthetic management should therefore be patient tailored. The potential for difficult airway, ventilation and the high incidence of spontaneous pneumothorax should be kept in mind[3].

CASE REPORT

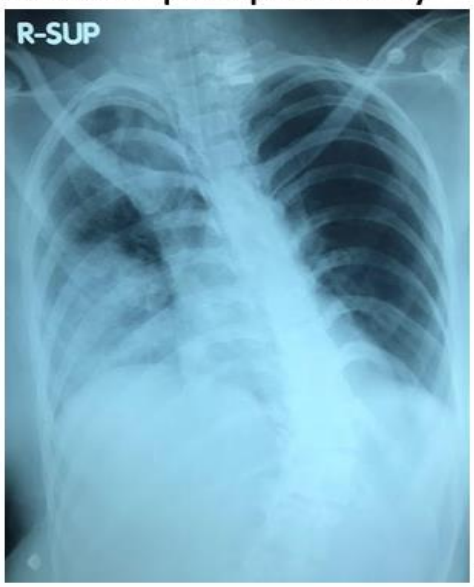
Our case report is of a 27-year-old primi, at a POA 37 weeks with a history of marfanoid features and bone marrow hypoplasia, who was admitted to the obstetric ward for the assessment and optimization for the delivery. The patient was poorly followed up and managed for her existing condition of bone marrow hypoplasia induced anemia and thrombocytopenia. Ultrasound scan of the fetus revealed severe growth restriction and an elective Caesarian section was planned.

Haematology opinion had suggested to transfuse six packs of platelets prior to the surgery.

The patient was seen by the anesthesia team only on the day of the surgery in the theatre. Her previous diagnosis of Marfanoid features and history of scoliosis were noted, but no chest X-ray was available. On examination she was found to have a systolic murmur, with a Saturation of 93% on air, no significant abnormality was found on respiratory examination. This low saturation was attributed to the supine position and splinted diaphragm due to gravid uterus. On airway assessment she had a mallampati of grade 3, Thyromental distance of > 6.5 cm, and good neck extension.

Fig.1

Immediate post op chest X-ray



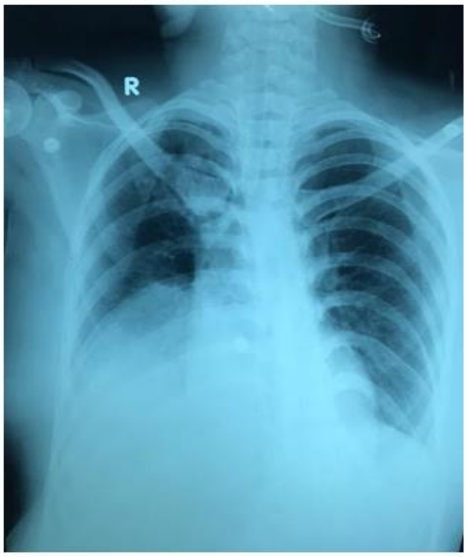
The anesthetist opted for general anesthesia due to her low platelet count (33,000/mcL) and the presence of significant scoliosis. Under basic monitoring, she was then slowly induced with Propofol (120mg) and Suxamethonium (100mg), after adequate pre-oxygenation. On direct laryngoscopy Cormack-Lehane grade 3 was visualized by the medical officer and the intubation was handed over to the consultant anesthetist. Bougie assisted intubation was then attempted and failed. The patient then showed signs of recovery from the neuro muscular block and mask ventilation was not adequate. Insertion of a supraglottic airway (SAD) was tricky as there was a possibility of airway bleeding due to existing coagulopathy which might complicate the situation further. She was then administered IV Atracurium (25mg) to optimize paralysis according to the plan C in Difficult Airway society guidelines. Difficult ventilation with high resistance was noted with no obvious upper airway obstruction even after adequate paralysis. She then started to rapidly desaturate, and her SpO_2 was maintained around 70-80% with 100% oxygen. Cannot intubate cannot oxygenate was declared (CICO). A second consultant anesthetist attended as requested. A quick examination revealed that there was no air entry to the right lung and lung collapse was suspected. Pneumothorax and bronchospasm were clinically excluded. FONA was delayed as the decision was made that the difficulty in ventilation was due to resistance to ventilation rather than a problem in the airway. The third Bougie assisted intubation attempted with size 7 ETT which was displaced into the esophagus. A fourth and successful attempt of intubation was carried out under bougie assistance with size 6.5 ETT. The lowest and the highest saturation noted were 50% and 85% respectively, during the entire sequence of events which took about 30 minutes. Her saturation then gradually improved from 85% to 98%. However, the right-side air entry remained almost absent. The anaesthetic and obstetric team decided to

proceed with LSCS as airway is secured and oxygenation was adequate. The intraoperative period was unremarkable with no significant bleeding. She was transferred post operatively to the SICU after evaluation of her conscious level.

Her immediate post-operative chest X-ray (Fig 1) shows R/S lower lobe collapse and marked thoracolumbar scoliosis. She was ventilated overnight. Her Chest X-Ray (Fig 2) improved after 24hours of positive pressure ventilation. She met the criteria for extubation on SICU day two and was extubated.

Fig.2

Chest X-Ray Day 02



A detailed retrospective evaluation was done. Our patient was first diagnosed with Marfanoid features with cardiac lesions at the age of 11, as an incidental finding. The other features that were suggestive were the arm span measure, chest deformity, hyper-flexible joint and a loud systolic murmur. She was found to have mild Left ventricular hypertrophy, myxomatous

degeneration of mitral valve and trivial Mitral regurgitation. A repeat 2D echo was done at the age of 23 and was found to have an aortic root measuring 1.2cm (Significant if >4 cm)[8,9], myxomatous MV with trivial MR, and mild asymmetrical septal thickening. At 25 years, she was admitted due to a viral infection and nasal bleeding and was found to have isolated thrombocytopenia. She was diagnosed with bone marrow hypoplasia with suppressed megakaryocytes after a Bone Marrow biopsy.

Patient's pregnancy was detected in April 2022 and thereafter followed by a cardiologist and hematologist for her respective conditions. Her family history was negative for Marfan syndrome.

Remarkable findings on examination were as follows[2]: General- She weighed 47kg. height of 146cm, an arm span to body height ratio of 0.8; pectus craniatum, wrist sign, pes planus, joint hypermobility, high arched palate and scoliosis with a Cobb angle of 60 degrees[7]. (Fig 3)
A loud systolic murmur, reduced air entry on the right, and a SpO2 of 97% on air. These features were compatible with Marfanoid features, but not the Marfan syndrome (MFS) This patient was discharged to the ward on SICU day three. Chest Physiotherapy was continued and her vitals were stable on discharge. The anesthetic events were clearly documented on a discharge card and the patient was educated on the anesthetic implications of her condition. A chest X-ray way taken one month after the incident. (Fig. 3 and 4)

DISCUSSION

MFS is a well described autosomal dominant disease which is caused by the mutation of FBN1 gene. In the above case report, the patient was labelled to have

Fig.3 One-month post-partum.
B/L lung fibrosis is more prominent

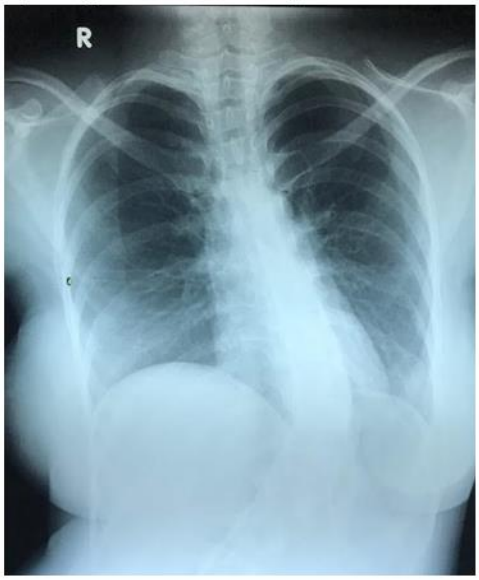
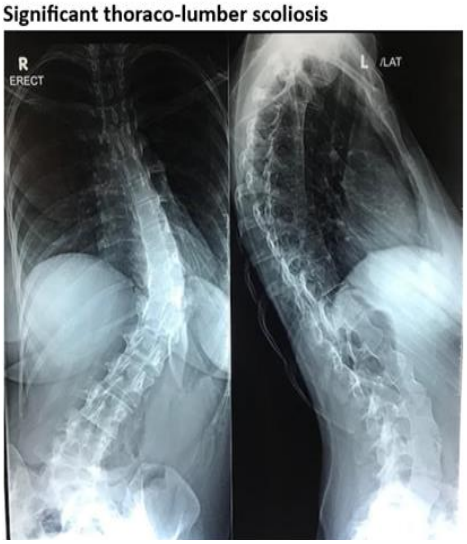


Fig.4 One-month post-partum
Significant thoraco-lumber scoliosis



Marfanoid features[5]. A productive perioperative care will begin with a thorough preoperative evaluation. The Revised Ghents Criteria (2010) is used as a guide for the diagnosis of the syndrome[1] (Table 1).

Table 1: Criteria for Marfan Syndrome Diagnosis from Revised Ghent Criteria
In the absence of family history
1. A0 (Z>/= 2) and EL = MFS 2. A0 (Z>/= 2) and FBN1 = MFS 3. A0 (Z>/= 2) and Syst (>7 points) = MFS 4. EL and FBN1 and known A0 = MFS
EL with or without systemic and with a FBN1 not known with A0 or no FBN1 = ELS A0(Z<2) and Syst (>/=5 with at least one skeletal feature) without EL = MASS MVP and A0 (Z<2) and Syst (<5) without EL = MVPS
In the presence of family history
5. EL and FH of MFS (as defined above) = MFS 6. Systemic (>/=7) and FH of MFS (as defined above) = MFS 7. A0 (Z>/=2 above 20 years old, >/=3 below 20 years) and FH of MFS= MFS
AO = aortic root diameter at the sinuses of valsalva above indicated z score or aortic root dissection, EL = Ectopia Lentis, ELS = Ectopia Lentis syndrome, FBN1= Fibrillin 1 mutation, FH = Family history, MASS= Mitral valve prolapse borderline (Z<2) aortic root dilatation, striae, skeletal findings phenotype ,MFS= Marfan Syndrome, MVPS= Mitral valve prolapse syndrome, Syst = Systemic score, Z= Z score

The primary concern to the anesthesia provider would be the potential for difficult intubation. A great proportion (59%) of the patients with MFS have high arched palate, 52% have TMJ dysfunction, and symptoms of subluxation were seen in 24% of the MFS population[2,3]. In this patient we were able to demonstrate a high arched palate which probably led to the primary hurdle during the induction i.e. the poor laryngoscopy view.

The presence of prognathism, high arched palate and crowded teeth may make the visualization of the larynx under direct laryngoscopy difficult. Our goal would be to establish airway with minimal C- Spine movement[3]. Inhalational induction can also be used with 100% oxygen maintaining spontaneous ventilation until bag mask ventilation can be demonstrated.

Tracheomalacia has been reported as a potential complication during and after anesthesia in patients with Marfan syndrome[3,6]. One also needs to pay attention to positioning of the patients as they are at risk of potential cervical spine (C1/2) ligamentous instability.

They are also predisposed to barotrauma secondary to bullae and apical blebs. The incidence of spontaneous pneumothorax in patients with MFS is as high as 4-15%[3]. The chest wall abnormalities that are seen in MFS are scoliosis and pectum excavatum. Scoliosis with a curvature of 60-100 degrees may lead to restrictive type lung disease[3], which can only be detected with pulmonary function testing, resulting in reduced total lung capacity.

Our patient presented with a pectus craniatum and scoliosis of 60 degrees (Fig.3).

Given the above complications, strategies such as low volume ventilation, increasing the inspiratory time, and the monitoring of exhaled tidal volume should be employed[3]. If there are any changes to the compliance or resistance, pneumothorax must be excluded[2].

Preoperative optimization of the pulmonary function is recommended. The use of Positive pressure ventilation may support avert post-operative respiratory insufficiency and failure in those with pre-existing pulmonary dysfunction.

Our patient was diagnosed to have Marfanoid features, which was not evaluated extensively by the anaesthesia team prior to the surgery as the patient was seen only on the day of surgery. Significant scoliosis and restrictive lung disease are possible with Marfanoid individuals, and pre-operative chest x-ray and lung function tests would be beneficial in the evaluation. This case highlights the presence of both difficult airway and restrictive lung disease which can give rise to very challenging intubating and ventilating conditions. In addition, patient had very low platelet count due to bone marrow hypoplasia and significant thoraco-lumber scoliosis (cob angle > 60%) which had led to make the decision to proceed with general anaesthesia even in the presence of predicted difficult airway. Deviation from the Difficult Airway Society (DAS) guidelines were consciously made by combined decision of both consultant anesthetists, as the situation was complicated by number of confounding factors.

The cardiovascular system is also frequently affected in patients with MFS, including aortic and pulmonary artery

dilation, valvular regurgitation, left ventricular dysfunction, and arrhythmias. Aortic root dilation is the most common cardiovascular manifestation of MFS[8,9]. Aortic root rupture and dissection remain the most life-threatening events in patients with MFS[8].

To summarize we have presented to you a case of Marfanoid features with an acute presentation to the anesthesia team and the fatal consequences that it may pose. The cardiovascular system of these patients presenting for anesthesia must be critically evaluated and optimized. The presence of any airway, chest and respiratory abnormality should raise red flags and a difficult airway must be anticipated. Preoperative evaluation is essential to identify anatomical and end-organ effects of MFS, as well as implications of therapeutic interventions. A written informed consent for publication was obtained from the patient.

REFERENCES

1.Loeys BL, Dietz HC, Braverman AC,Callewaert BL, et al The revised Ghent nosology for the Marfan syndrome. J Med Genet. 2010

2. Kamat S, Travasso B, Borkar D, et al. Anaesthetic considerations in a patient with Marfan's
syndrome for maxillary corrective osteotomy. Indian J Anaesth

3. MT Bösenberg & AT Bösenberg (2007) Anaesthesia for Marfan's Syndrome, Southern African Journal of Anaesthesia and Analgesia, 13:4, 15-19,

4. Singh SI, Brooks C, Dobkowski W. General anesthesia using remifentanil for Cesarean delivery in a parturient with Marfan's syndrome. Can J Anesth

5. Marfan syndrome: diagnosis and management; JCS Dean
Consultant and Honorary Reader, Department of Medical Genetics, Medical School, Aberdeen, Scotland

6. Tracheal Compression in Marfan Syndrome- Likurgos Kolilekas, Effrosyni Manali, Ioannis Kalomenidis, Ero Mouchtouri, et al

7.Adolescent Idiopathic Scoliosis: Radiologic Decision-Making K. ALLEN GREINER, M.D., M.P.H., University of Kansas Medical Center, Kansas City, Kansas

8. Choudhary SK, Goyal A. Aortic root surgery in Marfan syndrome. Indian J Thorac Cardiovasc Surg. 2019 Jun;35(Suppl 2)

9. Singh J, Wanjari A. Cardiac Complications in Marfan Syndrome: A Review. Cureus. 2022 Sep 30;14(9):e29