Case Report

Anaesthesia Management in a case of Marfans syndrome for Adenotonsillectomy: A case report

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Abstract

Background: Marfan syndrome is an Autosomal dominant multisystemic disorder involving mainly cardiac and respiratory system along with defect in connective tissue protein fibrillin 1. We hereby report successful respiratory and cardiac management of 10 years old male child with Marfans syndrome who presented with Mallampati Class IV, 4.5 cm of thyromental distance, left sided scoliosis, pectus excavatum with winging of scapula, pan systolic murmur, posted for adenotonsillectomy.

Keywords: Marfans syndrome; Adenotonsillectomy

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INTRODUCTION

Marfans syndrome was first described by Marfan in 1896. Children affected by the Marfan syndrome carry a mutation in one of their two copies of the gene that encodes the connective tissue protein fibrillin-1 (FBN 1). Affected individuals often are tall and slender, have arachnodactyly, scoliosis and either pectus excavatum, pectus carinatum or ectopia lentis in eyes. It has an estimated incidence of 2-3 per 10,000 inhabitants.[1] Fibrillin is an important component of microfibrils, and is essential for the integrity of both elastic and non-elastic connective tissue. Abnormal fibrillin alters the elasticity and tensile strength of connective tissue, particularly in areas where fibrillin is abundant such as the proximal aorta, zonule of the lens in the eye; in long bones and the skin.[2] Recent studies suggest that dysregulation of transforming growth factor beta (TGFbeta) signalling in lung, mitral valve and aortic tissues has been implicated in a subset of patients with Marfan syndrome.[3] Diagnostic criteria used by Ghent include a family history and involvement of at least two organ systems; or three organ systems if a mutation is suspected.[4] We herein report careful preoperative assessment combined with skilful anaesthetic technique can prevent cardiac and respiratory complications perioperatively.

CASE REPORT

A 10 year-old boy student, resident of Kolhapur to be posted for adenotonsillectomy, with chief complaints of recurrent episodes of cold and throat pain since one year. Tall (height-156 cm), thin (weight-34 kg) patient revealed typical features of Marfan syndrome; an arm span that was greater than his height (16 cm), narrow thin face, elongated fingers and toes (arachnodactyly), left sided scoliosis, pectus carinatum winging of scapula. On intraoral examination, revealed presence of high arched palate with MPC IV with 2 and 1⁄2 finger mouth opening and slight retrognathia which anticipated a potential difficult intubation, Systemic examination revealed pansystolic murmur heard in left parasternal area. Investigation revealed blood chemistry and haematocrit values were normal and CXR-Scoliosis of thoracic spine to the left with crowding of the ribs on the right, maintaining on low airway pressure to prevent the risk of pneumothorax. ECG suggestive of normal sinus rhythm with 2D echo-Mild AR, mild TR with LVEF-70%.

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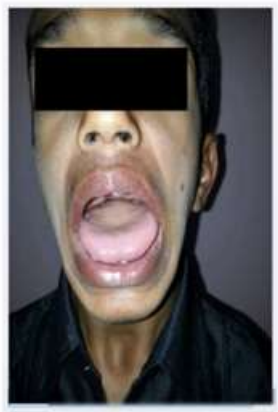

Figure 1

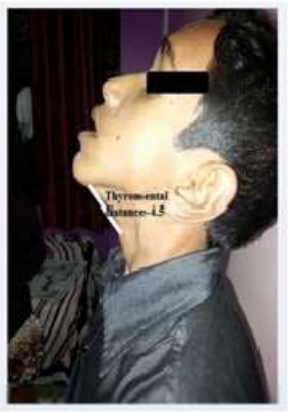

Figure 2

Figure 1: Mallampatti score; **Figure 1:** Shortened thyromental distance

Our plan A was to give general anaesthesia with Macintosh blade 3 with backup plan of reinforced laryngeal mask airway and emergency tracheostomy as plan B and C respectively in case of failed intubation with plan A. Adequate psychological preparation was insured to patient and parent as well. In operating room, ASA standard monitors were attached. Intramuscular injection of glycopyrrolate 4ug/kg given as antisialogogue. Preoxygenation was performed with 100% O2 for 5 minutes. Injection Fentanyl 2ugs/kg followed by injection propofol 2mg/kg given. Injection rocuronium 1mg/kg given. Intubation was done with Macintosh blade 3, bougie guided, with Cormack Lahane grade IIb using Endotracheal tube number 6. Confirmation of Endotracheal intubation done by capnography. Patient was maintained on oxygen, nitrous oxide and sevoflurane with maintaining on low airway pressure to prevent the risk of pneumothorax. Reversion of neuromuscular blockade was made with 2 mg of Inj. neostigmine and 0.6 mg of Inj. atropine. Patient was extubated after surgery uneventfully. Intraoperative and postoperative vitals were good without complications.

DISCUSSION

The diagnosis of classic Marfan's syndrome is based on presence of major criteria in at least two organ systems and involvement of a third organ system in absence of positive family history.[5] Majority of Marfan's syndrome patients are on beta blocker to slow the aortic dilatation by its negative inotropic and chronotropic effects.[6] It also has a beneficial effect in mitral valve prolapse (MVP) which is present in 80% patients of Marfan's syndrome.[7] In our case, we had mild Aortic regurgitation with tricuspid regurgitation. New diagnostic criteria for Marfans syndrome give more emphasis to aortic root aneurysm and ectopia lentis, with or without family history or positive FBN1 gene test leaving the systemic features on the backburner.[8] Proper diagnosis is crucial for the appropriate evaluation of the patient and to avoid predictable and potentially fatal complications such as rupture of an aortic aneurysm. On physical examination, one should be alert to signs of congestive heart failure. Echocardiography as indicated to assess the size of the aortic root and valvular function. Pulmonary function tests, with or without arterial blood gas should be considered if thoracic skeletal abnormalities are severe. As is anticipated, patients with Marfan's syndrome present with an increased incidence for ocular, cardio- vascular or musculoakeletal surgery. Major threats to life, severe cardiovascular complications, may occur at any time from infancy to the seventh decade. These complications include dilatation, dissection, or rupture of the aorta and severe regurgitation of the aortic or mitral valve.[9] Scoliosis is common and may occur at multiple sites along the thoracolumbar spine.[10] Even patients without a skeletal basis for pulmonary dysfunction demonstrate lower forced vital capacities than predicted, presumably as a result of earlier airway closure due to a lack of small airway elastic tissue support.[11] Proper positioning and limb support must be assured, considering the ligamentous hyperlaxity and increased risk of joint damage. The anaesthesiologist should be prepared for a potentially difficult intubation owing to factors related with high arch palate, retrognathia and ligamentous hyperlaxity that can lead to joint luxation during neck extension (cervical spine, temporomandibular joint).[12] Excessive traction at laryngoscopy should be avoided to prevent temporomandibular joint dislocation. Intraoperative cardiovascular monitoring is the core concern. Laryngoscopy should be as smooth as possible to prevent hypertension and subsequently increased risk of dissection. Regarding lung function, Marfans syndrome patients normally present with restrictive ventilatory defects, not only because of the underlying emphysema, but also due to the musculoskeletal changes that affect thoracic expansion.[13] Ventilatory pressures must be kept as low as possible to prevent barotrauma and reduce the risk of pneumothorax. Spontaneous pneumothorax are a frequent occurrence. The potential for the development of a pneumothorax must always be borne in mind during anaesthesia, particularly with positive pressure ventilation. Changes on AV conduction or perioperative dysrhythmias are common small arteries supplying the sinus and AV nodes were especially involved, probably explaining the higher frequency of arrhythmias and conduction disturbances.[14] Tracheomalacia has been reported as a potential complication.[15] As for intraoperative fluid therapy, the primary goal is to maintain blood volume in order to decrease the chances of aortic and/or mitral valve prolapse.[1]

CONCLUSION

Pre-existing cardiovascular disease and the potential for acute cardiovascular and respiratory complications in patients with Marfans syndrome demand careful preoperative assessment and the use of skilful anaesthetic technique to avoid fatal complications. Blood pressure control is the central component of perioperative management. The risk of perioperative morbidity and mortality, including unexplained death, is high.[17] Multidisciplinary approach is key. Whatever the anaesthetic technique, the primary consideration is the avoidance of sudden increases in myocardial contractility, producing an increase in aortic wall tension. Beat to beat monitoring by an intra-arterial cannula is helpful, but should be performed carefully in these patients who may be at increased risk from morbidity attributable to weakened arterial walls.

DECLARATION OF PATIENT CONSENT

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for his images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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