Case Report

Emergent airway management in a case of fibrodysplasia ossificans progressiva

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Abstract

Fibrodysplasia ossificans progressiva (FOP), or Stone man syndrome, is rare and one of the most disabling genetic conditions of the connective tissue due to progressive extraskeletal ossification. It usually presents in the first decade of life as painful inflammatory swellings, either spontaneously or in response to trauma, which later ossify and lead to severe disability. Progressive spinal deformity including thoracolumbar kyphoscoliosis leads to thoracic insufficiency syndrome, increasing the risk for pneumonia and right sided heart failure. We present the airway management in a 22-year-old male, diagnosed with FOP with severe disability, who required urgent airway intervention as a result of respiratory failure from pnuemonia. Tracheostomy triggers ossification and consequent airway obstruction at the tracheostomy site and laryngoscopy triggers temporomandibular joint ankylosis. Therefore, awake fiber-optic endotracheal intubation is recommended in these patients. Use of an airway endoscopy mask enabled us to simultaneously maintain non-invasive ventilation and intubate the patient in a situation where tracheostomy needed to be avoided.

Key words: Airway endoscopy mask, airway management, fibrodysplasia ossificans progressiva

Introduction

Fibrodysplasia ossificans progressiva (FOP) is one of the most disabling genetic conditions of the connective tissue in humans due to progressive extra skeletal ossification, either spontaneously or in response to trauma. It is a rare disorder with world-wide prevalence of one in two million. Genetic transmission is autosomal dominant, but most cases occur as a result of a spontaneous mutation.[1] Cardiorespiratory failure from thoracic insufficiency syndrome (TIS) due to ossification of thoracic muscles is the most common cause of mortality in the third or fourth decade of life.[2] Although there are a few reports describing anesthetic management of FOP patients in elective situations,[3,4] there is a dearth of literature regarding emergent scenarios. We report an emergent difficult airway management in a patient with advanced FOP where surgical airway should be avoided.

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Case Report

This was a case of a 22-year-old male patient, weighing 18 kg, diagnosed with FOP, presenting with severe pneumonia progressing to respiratory failure. His clinical features included severe kyphoscoliosis with restrictive lung disease, necessitating non-invasive bi-level positive airway pressure (BIPAP) therapy (settings of 5/4 cm H_2O), critical temporomandibular joint (TMJ) ossification causing restricted mouth opening (<1 cm) and consequent severe malnutrition from poor feeding, chronic gastro esophageal reflux, multiple musculoskeletal deformities, including multiple contractures of arms and legs and chronic pain syndrome. His medications included baclofen, fentanyl patch, oxycodone, omeprazole, metoclopramide and albuterol and fluticasone/salmeterol inhalers.

Physical examination revealed an emaciated patient weighing 18 kg non responsive to verbal commands with a Glasgow coma scale of 11. His neck was fixed in moderate flexion. O_2 saturation ranged between 88% and 96% despite oxygen administration (FiO_2 100%) with BIPAP support of 18/5 mm Hg. Arterial blood gas showed severe respiratory

acidosis, with pH of 6.98, $PaCO_2$ 136 mm Hg and PaO_2 99 mmHg. The chest X-ray was difficult to interpret due to extensive kyphoscoliosis, but did reveal right sided tracheal deviation [Figure 1]. Patient was kept nil *per os* and maintained on intravenous fluids. An anesthesia consultation was initiated to evaluate the patient for tracheal intubation and positive pressure ventilation.

In the operating room, with an ENT team available to perform an emergent tracheostomy should that be absolutely necessary, an airway endoscopy mask (VBM mask; # 3, child; hole 3 mm; ref: 30-40-333) [Figure 2] was used to intubate the trachea with a fiber-optic intubation videoscope (Olympus LF-V) while maintaining him on pressure support of 18 mm Hg and positive end-expiratory pressure of 5 mm Hg with 100% oxygen through the anesthesia machine.

The patient was positioned with pillows and blankets under the right thorax and head to get optimal access to the airway and prevent any ossification from pressure injuries. Electrocardiogram (ECG), pulse oximetry and non-invasive blood pressure monitoring were instituted. An intravenous bolus of 10 mcg dexmedetomidine was infused over 5 min and 10 mg ketamine was administered intravenously. Nostrils were prepared with xylometazoline drops and dilated with a 20 mm nasopharyngeal airway. Fiber-optic bronchoscopy was performed through the airway endoscopy mask to determine the degree of visualization of the glottic aperture. The vocal cords were visualized with some difficulty. There appeared to be some subglottic stenosis. A further bolus of 10 mg of ketamine was administered intravenously. The bronchoscope was removed and reintroduced with a 5.0 mm cuffed microlaryngeal tube threaded over it. The endotracheal tube was carefully guided through the vocal cords. Rocuronium 20 mg was administered intravenously after intubation. Brownish secretions, which could have been due to aspiration, were noted in the tube. The patient remained hemodynamically stable and oxygen saturations were maintained between 82% and 96% during the procedure. The endotracheal tube appeared to be endobronchial but could not be withdrawn, probably due to airway edema. Dexamethasone (8 mg) was administered and the tube was pulled back into the trachea a few hours later in the intensive care unit.

The patient was discharged with the endotracheal tube and home ventilation 36 days after intubation. Tracheostomy was not performed.

Discussion

We describe the airway management and complications in a very challenging scenario of advanced FOP in a patient who, by chronological age, belongs to the adult population but was the size of a 4-5 year old child.

FOP presents in the first decade of life as painful and highly inflammatory soft-tissue swellings, either spontaneously or in response to trauma, which then progress to ossification. Ectopic ossification occurs in tendons, ligaments, joint capsules and muscles, causing immobilization.[1] One of the most common complications is extra-articular ankylosis of the TMJ leading to severe disability, resultant eating difficulties and poor oral hygiene.[5] Neck stiffness, due to cervical spine anomalies, is an early presentation. Ossification of intercostal muscles and paravertebral muscles and progressive spinal deformity, including thoracolumbar kyphoscoliosis, lead to TIS causing pneumonia and right-sided heart failure.[6] Ossification of cardiac muscle has not been described, but cardiac connective tissue may be involved, leading to ECG abnormalities including right bundle branch block, T-wave inversion, left axis deviation with ST segment changes and supraventricular tachycardia.[6]

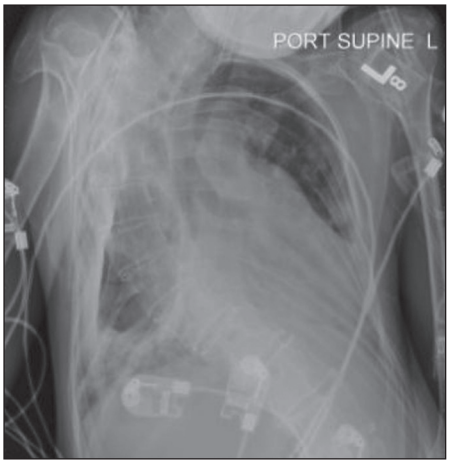

Figure 1: Chest X-ray of patient with fibrodysplasia ossificans

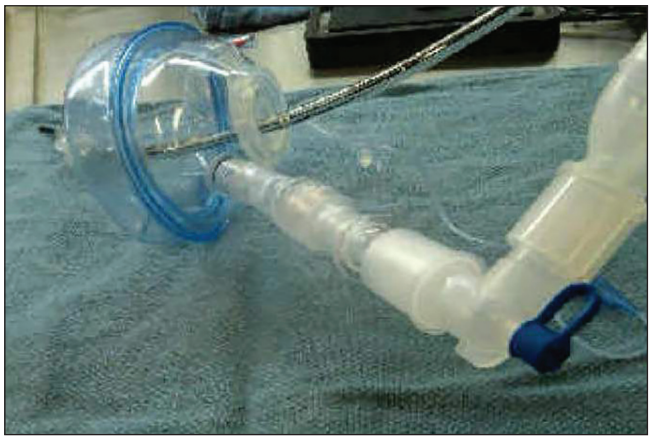

Figure 2: Airway endoscopy mask (VBM mask; # 3, child; hole 3 mm; ref: 30-40-333)

Pre-operative evaluation should include chest X-rays, ECG and if possible, spirometry to assess cardiopulmonary function. Cervical spine status and TMJ should be assessed as fusion of the cervical vertebrae and TMJ ankylosis are common features.[7]

Currently corticosteroids, non-steroidal anti-inflammatory drugs, cyclo-oxygenase-2 inhibitors, leukotriene inhibitors (montelukast) and mast-cell stabilizers have been used in the treatment of FOP and flare-ups.[8] Minor trauma, such as intramuscular or intraoral injections, venepuncture, muscle fatigue and blunt muscle trauma from bumps or bruises, commonly leads to episodes of explosive and painful new bone growth.[8]

Laryngoscopy and maneuvers like jaw thrust should be avoided to prevent stretching of the TMJ and consequent ossification. Previous reports of general anesthesia recommend awake fiber-optic endotracheal intubation.[7] Succinylcholine could be contraindicated due to disuse atrophy. Tracheostomy and transtracheal injections should be avoided because of possible ossification in the incision and resultant airway obstruction.[9] However, there is a recent report of emergent tracheostomy in a child with FOP.[10]

Airway endoscopy mask was chosen for simultaneous noninvasive ventilation and fiber-optic intubation. It is a disposable mask with an opening on the mask covered with a silicone membrane and a ventilation port to the side of the mask [Figure 2]. A fiber-optic bronchoscope can be passed through the opening in the silicone membrane, which maintains an effective seal, whereas ventilation can be simultaneously maintained through the ventilation port. Da Conceiçao et al., have described the use of an airway endoscopy mask for fiber-optic intubation in adults.[11] Dexmedetomidine and ketamine provided sedation without cardiorespiratory compromise. We used the microlaryngeal tube since a 5.0 cuffed nasal endotracheal tube was not available at that time. Every effort should be made to avoid situations that may put the patient at risk of new localization of heterotrophic ossification.

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