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CLINICAL REPORT

Anesthetic management for foramen magnum decompression in a patient with Morquio syndrome: a case report

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Abstract Morquio syndrome is a hereditary mucopolysaccharide disorder presenting with an abnormality of the craniocervical junction from childhood. We describe an adult patient who presented with Morquio syndrome who had subglottic narrowing of the airway, restrictive pulmonary disease, and mild mitral regurgitation and trivial aortic regurgitation. The anesthetic management of this patient for atlantoaxial stabilization is presented.

Keywords Mucopolysaccharidosis · Difficult airway · Fiberoptic intubation · Cervical myelopathy · Atlantoaxial dislocation

Introduction

Morquio syndrome, mucopolysaccharidosis IV, is an autosomal recessive disorder of connective tissue characterized by disproportionate dwarfism and associated with skeletal deformities, particularly genu valgum, pigeon chest, and lumbar gibbus. Prominent maxilla, short and unstable neck, widely spaced teeth and receding mandible are the reported causes of difficult airway in these patients [1]. We report a case of Morquio syndrome with subglottic narrowing, restrictive pulmonary function, mild mitral regurgitation and trivial aortic regurgitation that was managed successfully for foramen magnum decompression.

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Case report

A 20-year old woman with an established diagnosis of Morquio syndrome presented with neck pain, difficulty walking and getting up from a squatting position, and limb stiffness for 2 months. She was born to consanguineous parents and had disproportionate limb and facial features and a history of snoring. She was 112 cm tall, weighed 21 kg, and had a head circumference of 52 cm, neck circumference of 18 cm, and arm span of 101 cm. Her heart rate, blood pressure, and respiratory rate were normal. Her limbs were short. She had a fixed flexion deformity of the shoulders and elbow, wrist, and knee joints. She had short and stubby fingers with thumb in the same plane as the other fingers, bilateral club foot with hallux valgus deformity, and thoracic kyphoscoliosis. She had grade 4/5 muscle power with increased tone in the lower limbs. Romberg's sign was positive. Airway examination revealed a large tongue with a Mallampati grade 3, an interincisor gap of 3 cm, a thyromental distance of 4 cm, receding chin (Fig. 1), and prominent teeth. Her hemogram, renal and liver function tests, and blood glucose were within normal limits; electrocardiogram (ECG) showed a right axis deviation. An echocardiogram revealed mild mitral regurgitation and trivial aortic regurgitation; pulmonary artery systolic pressure was 28 mmHg and the ejection fraction 59%.

Her chest radiograph showed prominent bronchovascular markings with wide ribs (Fig. 2) and a subglottic narrowing of tracheal air shadow (Fig. 3). An X-ray of the

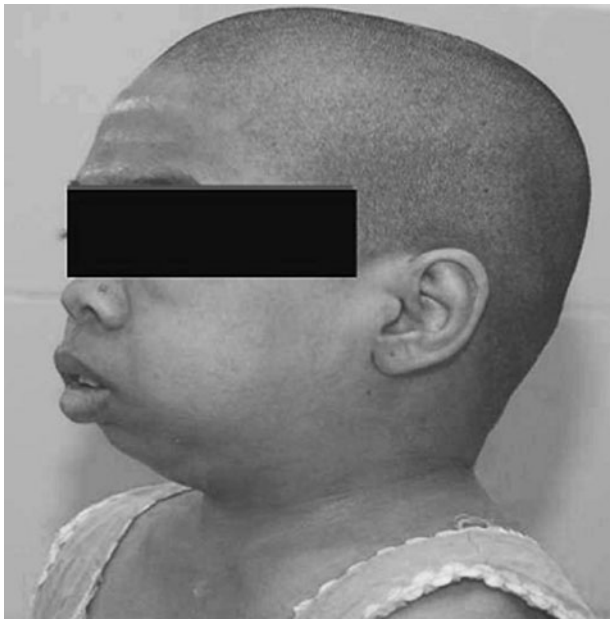

Fig. 1 Lateral view of the patient. Note short neck, receding mandible

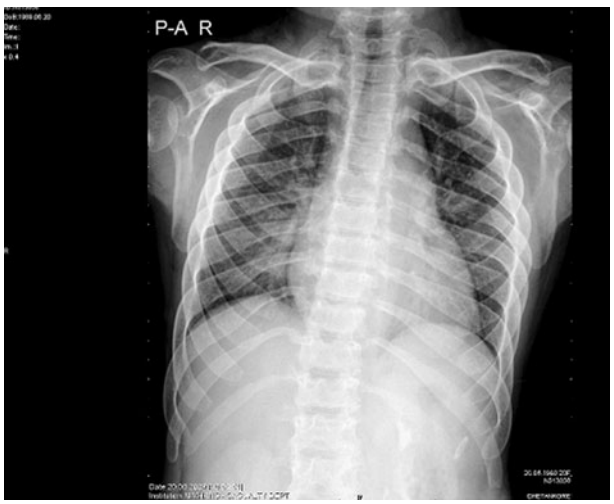

Fig. 2 Posteroanterior chest X-ray. Note wide ribs

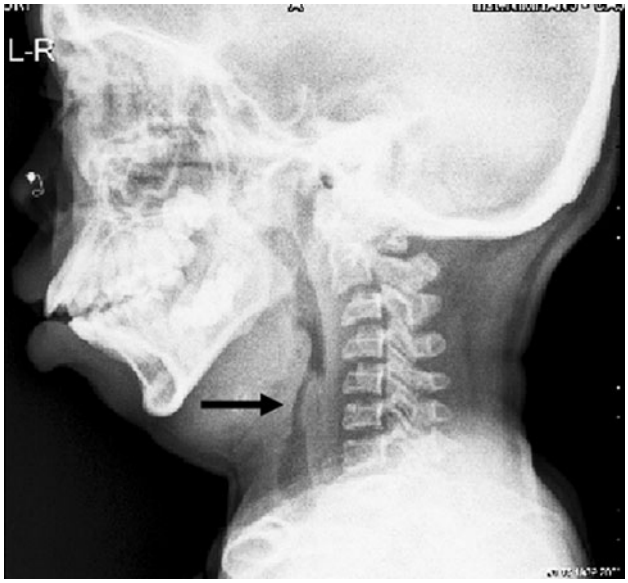

Fig. 3 Lateral X-ray of the neck showing subglottic narrowing (*arrow*)

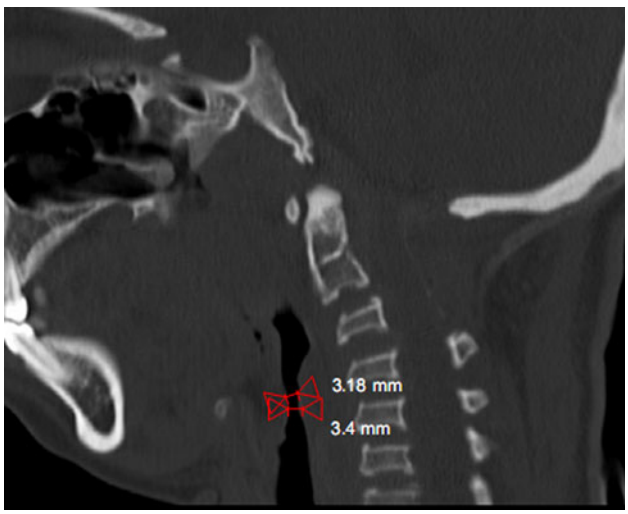

Fig. 4 Computed tomography (CT) of the neck, sagittal section. Note airway narrowing at the C4–C5 level. Measurements were 3.18 mm (*top*) and 3.4 mm (*bottom*)

cervical spine revealed narrowing of the vertebral canal at the craniovertebral junction, with an effective canal diameter of 8 mm (normal value > 18 mm). Computed tomography (CT) of the spine confirmed C_1 arch stenosis with dysplastic odontoid. CT of the neck revealed airway narrowing at the C4–C5 level (Fig. 4).

The patient's pulmonary function tests (PFT) showed severe restrictive abnormality with a forced vital capacity (FVC) of 0.73 L [60% of predicted value (1.2 L)], peak expiratory flow rate (PEFR) of 0.6 L/s [21% of predicted value (2.8 L/s)], forced expiratory flow ($FEF_{25–75}$) of 0.13 L/s [7% of predicted value (1.9 L/s)], and maximal voluntary ventilation (MVV) of 13.8 L/min. Anticipating a difficult airway, fiberoptic intubation was attempted after premedication with midazolam 0.5 mg, fentanyl 10 μg, and glycopyrrolate 0.1 mg, and airway nebulization with 3 ml of 4% lidocaine with 1 ml 0.9% normal saline for 15 min through a mouthpiece using a jet nebulizer in the sitting position. The posterior pharyngeal wall was sprayed with 10% lidocaine (two puffs). Monitoring during the procedure included ECG, noninvasive blood pressure, pulse oximetry, temperature, and capnogram.

Initially, an adult-size fiberoptic bronchoscope (FOB) (Pentax, model-13RBS, 4.2 mm OD) was used to attempt tracheal intubation. Due to the short stature of the patient

and a narrow subglottic area, a 28-Fr (6.5 mm ID; 9 mm OD) cuffed reinforced endotracheal tube (ETT) was chosen. The epiglottis and vocal cords could be seen, but the ETT could not be passed below the vocal cords. As a smaller size ETT could not be threaded over the adult size FOB, and a smaller size reinforced ETT would have increased the airway resistance, intubation was achieved with a 5 mm ID; 6.9 mm OD (SIMS Portex Limited, UK) polyvinyl chloride (PVC) uncuffed ETT using a pediatric FOB (3.5 mm OD). The patient tolerated the procedure well.

Anesthesia was induced with thiopentone and maintained with sevoflurane and intermittent doses of fentanyl. Her lungs were ventilated with oxygen–air mixture at a respiratory rate of 12/min and a tidal volume of 180 ml using atracurium for muscle relaxation. The patient was positioned prone for surgery, taking care to avoid excessive movement of the neck. The patient's head was fixed in the neutral position with a Mayfield clamp. The intraoperative course was uncomplicated, with heart rate and blood pressure maintained within 15% of her baseline levels, end tidal carbon dioxide (CO_2) between 32 and 36 mmHg, and the peak airway pressure at 20 cm water (H_2O). At the end of the procedure, her trachea was extubated. Her postoperative course was uneventful.

Discussion

Mucopolysaccharidoses represent a group of hereditary disorders caused by defects in the degradation of mucopolysaccharides and resulting in accumulation of incompletely catabolized mucopolysaccharides in the connective tissues, especially bone, brain, liver, blood vessels, skin, cartilage, airways, heart valves, and cornea. Morquio's syndrome, otherwise called mucopolysaccharidosis IV, results from accumulation of keratan sulfate due to the deficiency of *N*-acetyl-galactose-6 sulfate sulfatase [2]. These children are normal at birth but demonstrate spinal dysplasia within 12–18 months. Abnormality at the craniocervical junction is almost universal, with hypoplastic odontoid, atlantoaxial instability (AAI), and, in some cases, severe cervical cord compression [3, 4]. These patients develop quadriparesis at a young age because of AAI. Our patient developed AAI acutely in adulthood following a minor injury, possibly from the worsening of a pre-existing asymptomatic AAI.

Patients with Morquio's syndrome have preserved higher mental functions in comparison with other mucopolysaccharidoses [2]. This helped us in performing awake fiberoptic intubation with minimal sedation. We preferred the FOB technique over other techniques, such as a specifically designed plaster bed for neck fixation and angulated video intubation laryngoscope [5, 6], as the patient had an unstable short neck and abnormal facial features, which would have rendered direct laryngoscopy impossible. Moreover, a large head with a short neck and trunk might aggravate cervical instability in dwarfed patients [7]. A superior laryngeal nerve block or a transtracheal block was not performed, as they require neck movement to identify anatomical landmarks.

The severe subglottic narrowing seen in our patient has not been described in earlier reports. The narrowing could be due to accumulation of mucopolysaccharides in laryngeal and tracheobronchial cartilages. The subglottic narrowing prevented us from placing even the smallest reinforced ETT using an adult FOB. The plan to use a smaller-sized reinforced ETT over a pediatric FOB was abandoned considering the resistance to ventilation. We could successfully use an uncuffed PVC tube, as the head position was maintained neutral during surgery.

Our patient had a history of snoring but no evidence of pulmonary hypertension, which is generally reported in patients with Morquio syndrome with obstructive sleep apnea. Respiratory mechanics may be compromised from airway obstruction, pectus deformities, mechanical distortion of the thorax, or deposition in the tracheobronchial tree [2]. Even though our patient had restrictive abnormality on PFT, she was comfortable at rest. Her PFT values, when looked at in isolation, were suggestive of restrictive abnormality. However, because of dwarfism, her height and weight might not correlate, and hence the lack of correlation between the measured values of PFT and values as predicted by the reference nomogram. Therefore, caution should be exercised in interpreting pulmonary function status from spirometry values in these patients [8].

Difficulty in artificial ventilation caused by chest deformity, reduced chest-wall compliance [8], and airway collapse [9] described in earlier reports were not experienced by our patient. Involvement of other organ systems is also seen in patients with Morquio syndrome [10]. The right bundle branch block and trivial mitral and aortic regurgitation did not pose any serious problems in our patient.

In summary, we present a case of Morquio syndrome with difficult airway and subglottic narrowing, severe restrictive pulmonary dysfunction, and mild cardiac abnormalities managed successfully for a foramen magnum decompression. To our knowledge, there are no other case reports of anesthetic management of atlantoaxial stabilization in a patient with Morquio syndrome and subglottic narrowing.

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