CASE REPORT OPEN ACCESS

Challenges In the Management of Acute Airway Obstruction in A Toddler with Cervical Macrocystic Lymphatic Malformation (Cystic Hygroma): A Case Report

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ABSTRACT

We report an 18-months-old female toddler with macrocystic lymphatic malformation (cystic hygroma) of cervical region who presented with severe airway compromise. Considering the increase in the size of swelling and its effect airway on urgent intervention was required. High-flow supplemental oxygen was provided by a non-rebreathing mask. However, severe respiratory acidosis and hypoxemia persisted. As imminent respiratory failure was feared due to airway obstruction a need to secure definitive airway was identified. Attempts at endotracheal intubation failed. Decision was taken to perform needle cricothyroidotomy but anatomical landmarks could not be appreciated. Tracheostomy was then performed which was a challenge as trachea was difficult to identify. This case report highlights the steps taken in the management of compromised airway for general information of readers.

Key words *Macrocystic lymphatic malformation, Cystic hygroma, Tracheostomy, Respiratory failure, Child.*

INTRODUCTION:

Macrocystic lymphatic malformation, previously named as cystic hygroma, is one of the lymphatic malformations (LMs). It is a benign condition with an incidence of approximately 1/6000 live births.[1,2] Different theories have been proposed as to the origin of this condition from sequestration to improper anastomosis of lymphatic vessels with vascular system.[3] LMs are usually present at birth and most commonly found in the cervical and axillaryregions.[2] Rapidly enlarging cervical macrocystic lymphatic malformations cause acute airway obstruction depending upon the size and location of the cysts.[4] We present a patient with a huge right sided cervical mass that was asymptomatic in infancy but then caused acute airway obstruction with the growth of the child.

CASE REPORT:

An 18-month-old female was referred from another city with the complaints of acute respiratory distress for two days secondary to large neck swelling. The child had swelling on the right side of the neck since birth which was initially small in size. Consultation was made with a general practitioner but no intervention was advised. Four weeks prior to presentation the girl was admitted to a nearby hospital with the complaints of fever, cough, and running nose. During this period the neck swelling gradually increased in size. According to the attendant, the girl developed acute respiratory distress two days before her presentation to our hospital.

On arrival in the ER child had severe respiratory difficulty with nasal flaring and subcostal and intercostal recessions. Her respiratory rate was 60 breaths per minute and heart rate 148 beats per minute. There was a large multiloculated swelling in the right side of the neck involving the mandible, floor of the mouth, anteriorly crossing the midline to the left and posteriorly extending to the occipital region. The lower limit of the swelling was just above the supra-sternal notch. The swelling was tense. However, there were no signs of inflammation. It was not possible to palpate the trachea due to overlying swelling. However, thyroid cartilage could be felt deep into the swelling and was found to shift towards the left side. The cxygen saturation in room air was 90%. High-flow supplemental oxygen was provided by a non-rebreathing mask. Baseline investigations and arterial blood gas analysis (ABG) were done. The report was suggestive of respiratory acidosis and hypoxemia. Considering the chances of imminent respiratory failure due to airway

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obstruction and possible pneumonia a plan was made to secure a definitive airway to support oxygenation and ventilation.

The operation theater was prepared with age and weight-appropriate airway equipment, including a face mask, LMA, bougie, endotracheal tube (ETT) of size 4.0mm and 4.5mm, and video laryngoscope. The senior anesthesiologist with his team were on board. The instrument trolley was on standby for emergency needle cricothyroidotomy and tracheostomy. The patient was then shifted to the operation theater and pre-oxygenation was carried out for three minutes with 100% oxygen and maintaining a saturation of 99%. The child was kept in a lateral position. After oxygenation propofol was given and a laryngoscopy performed. It was not possible to visualize the vocal cords. A decision was made to aspirate some of the cystic fluid under anesthesia with continuous ventilation via a mask in a controlled manner. With a syringe aspiration of fluid was done from the anterior part of the cystic swelling that was suspected to cause the displacement of the trachea. Following aspiration a slight reduction in the size of the swelling was noticed. Another attempt to pass ETT was made but this time child went into bradycardia and CPR was initiated. Injection atropine was also given. After improvement of the heart rate plan was made for immediate needle cricothyroidotomy or tracheostomy depending upon the situation.

The procedure started with an incision made left of the midline as the trachea was pushed to one side. A firm structure was felt which was suspected to be the thyroid cartilage. The anterior neck muscles were dissected. However, due to the distorted anatomy resulting in significant displacement, it was not possible to visualize the trachea. Another attempt was made to aspirate the cysts following which the trachea could be identified below the swelling. Manual displacement of the neck mass was made away from the trachea which was then opened in midline vertically. A tracheostomy tube of size 4.0mm was passed and the balloon inflated. After clinical assessment of the proper placement of the tube the airway was secured. The patient was then shifted to the surgical intensive care unit for mechanical ventilation (Figure I).

The postoperative course remained stormy. Invasive ventilation continued and parameters had to be adjusted multiple times as the patient was unable to maintain the ventilation. Meanwhile, the general care continued with IV fluids, IV antibiotics, and antipyretics. X-ray chest showed features of bilateral consolidation. However, despite all the measures patient passed away 48-hours after tracheostomy.

DISCUSSION:

Lymphatic malformations are usually present at birth. Number of these lesions can be diagnosed on antenatal ultrasound.[5] In our patient, the malformation was apparent at birth. The LM can involve any part of the body but are most commonly found in the cervical and axillary regions.[6] Our patient had lesion on the right side of neck. Large swelling in cervical region can result in obstructed labor during delivery and feus is also at a risk. In such cases the delivery can be attempted through an ex-utero intrapartum treatment (EXIT) procedure.[7]

Fig I: Child with huge lymphatic malformation of cervical region

Although LMs have benign clinical presentation, however number of complications are reported when they get infected or intra-cystic hemorrhage occur. This leads to sudden increase in size of the lesion. This results in respiratory distress if cervical region is involved as happened in our patient.[8] Antibiotics should be started early to control the infective process.[9] In our patient though treatment of pneumonia was started early but sudden increase in the size of the LM lead to the tracheal obstruction and condition worsened. At this point surgeon and anesthesiologist should be prepared for various interventions to secure definite airway.[10] Intubation in such a situation requires drugs (drug assisted intubation - DAI) and a competent anesthesiologist. Same was tried in our patient, however even after two attempts it failed as vocal cords could not be visualized.

Video-assisted laryngoscopy may help in such a situation. Partial aspiration of the cysts might facilitate intubation that we tried in our patient. Cricothyroidotomy or tracheostomy were also difficult

in our patient as swelling covered anterior part of the neck. A desperate attempt for tracheostomy was made as patient was revived following cardiac arrest. With manual pushing of the swelling to one side and deep palpation helped in identifying tracheal rings and tracheostomy was made. However, our patient could not survive, probably due to hypoxic brain injury even when ventilator support and other care was provided.

CONCLUSION:

Macrocystic lymphatic malformations usually have a benign course. Different treatment options are available which must not be delayed because potential complications specially in cervical region can be threatening. Neck is a confined space within deep fascia encircling the important vital structures. It results in pressure on airway and distort the anatomy. Early referral to a tertiary care center before the condition deteriorates may help in salvaging such children.

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All authors were involved in the management of the child. They conceived the idea of reporting the case. All were involved in literature review, manuscript writing and revision. All authors are responsible for the content of the manuscript.

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Ethics statement. The consent of the parents was taken to report this case for educational purposes. Pictures are produced with the permission of the parents. Confidentiality is maintained while reporting the case.

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