Case Report

CYSTIC HYGROMA: ANAESTHETIC CONSIDERATIONS IN EMERGENCY EXCISION

I.M. Chukwu[a], J. Ojebo[a], F. O. Omosofe[b], N.P. Edomwonyi[c]
a.Department Of Anaesthesia/Intensive Care, Irrua Specialist Teaching Hospital, Edo State, Nigeria
b. Department Of Anaesthesia/Intensive Care, Irrua Specialist Teaching Hospital, Edo State, Nigeria
c. Department Of Anaesthesia/Intensive Care, University Of Benin Teaching Hospital, Edo State, Nigeria

ABSTRACT

This is a case report of an 8-month-old male who presented with right huge infected haemorrhagic cystic hygroma with airway obstruction and pharyngotonsillitis. Right cystic hygroma was diagnosed at 3 months of age but was managed conservatively until he fell from a height and traumatized the tumour which rapidly increased in size and resulted in airway obstruction necessitating emergency excision. The excision was done under general anaesthesia with intermittent positive pressure ventilation and muscle relaxation. He was subsequently transferred to the intensive care unit with the endotracheal tube in-situ and mechanically ventilated for 3 days. The patient was thereafter extubated and discharged to the ward on the 5th day in good clinical condition.

Keywords: cystic hygroma, emergency excision, difficult airway, preparedness.

NigerJmed 2020: 166-168

INTRODUCTION

Cystic hygroma is a congenital cystic tumour caused by a malformation of the lymphatic system leading to obstruction between the lymphatic and venous systems and accumulation of lymph. It is commonly seen in the neck, and less often, in the axilla, mediastinum, groin and retroperitoneum. Diagnosis can be made prenatal, at birth, infancy or even at adulthood. Incidence is between 1: 6000 – 12,000 live births. About 90% of the cases are diagnosed within 2 years of age.[1] Treatment is ultimately surgical. Some non-surgical treatments include repetitive suction, intralesional administration of sclerosant such as bleomycin, and radiotherapy.[2,3] The anaesthetist is faced with airway challenges perioperatively. The patients usually present with airway obstruction in the presence of a huge neck mass, and there could be difficult airway control during induction of general anaesthesia. After endotracheal intubation, there may be accidental extubation during positioning. Most of these patients also develop post-operative respiratory obstruction due to inflamed airway and distorted tongue anatomy. This should be anticipated, and the patient will benefit from close monitoring after surgery. The case of cystic hygroma being reported was previously managed conservatively for 4 months until the mass was traumatized and rapidly increased in size. The patient developed sudden marked airway obstruction, which necessitated emergency excision. There was a co-existing pharyngotonsillitis, and the cyst was also infected. Postoperative airway oedema was anticipated, and the plan was instituted to maintain airway patency after surgery.

Correspondence to:
Prof. N.P Edomwonyi
Department of Anaesthesia/ Intensive care,
University of Benin Teaching Hospital,
Edo State, Nigeria
E-mail Address: nosaphilo@gmail.com
+234 8153357133

CASE REPORT

An 8-month-old male infant weighing 8.1kg who presented at Irrua Specialist Teaching Hospital with right neck swelling of 5 months duration; noisy and difficulty with the breathing of 2 days; fever and reduced feeding of 1-day durations. He was diagnosed 4 months prior to presentation as a case of right cervical cystic hygroma with bronchopneumonia and has had conservative management for septic and obstructive symptoms and congestive cardiac failure. His condition worsened after he fell from a height two days before presentation and developed a rapid increase in the size of the neck mass with subsequent severe respiratory embarrassment.

On examination, he was acutely ill-looking, dyspneic with a respiratory rate of 54 cycles per minute, flaring of alae nasi, intercostal and subcostal recessions with inspiratory stridor. He was febrile (axillary temperature of 38.7^{0}C), and heart rate was 160 beats per minute. Neck mass was 9cmx10cm on the right extending from below the right tragus and midline of the submandibular region down to the thoracic inlet with extension to the left side of the neck. No demonstrable intrathoracic extension. The surface of the mass was smooth, soft, tender, fluctuant and did not transilluminate. Oral examination revealed intra-oral involvement, and inflamed tonsils and pharynx. A diagnosis of right-sided, infected and haemorrhagic huge cystic hygroma with upper airway obstruction and

pharyngotonsillitis was made. He was immediately prepared for emergency excision biopsy.

Full blood count panel revealed Haemoglobin concentration of 9.6g/dl, Packed Cell Volume of 29.9%, white blood cell count of 13,000 cells/mm^3, Neutrophil/lymphocyte of 69.9%/25.6% respectively and platelet count of 335,000 cells/mm^3. Serum electrolytes, urea and creatinine results showed Na^+ (138mmol/L), K^+ (4.3mmol/L), HCO^{-3} (27mmol/L), and urea (14mg/dl). The patient blood group was determined and crossmatched with the donor blood. Chest radiograph showed tracheal compression and deviation to the left, but no intrathoracic extension.

The parents of the patient were counselled on the potential difficult airway and the possibility of postoperative admission into the intensive care unit (ICU). He was transferred to the theatre. Difficult airway tray was prepared, and Ear, Nose and Throat (ENT) surgeon was on standby for emergency tracheostomy should endotracheal intubation fail. The nasogastric tube was inserted and suctioned to drain the stomach content.

Monitors were attached including a pulse oximeter, electrocardiogram leads and precordial stethoscope, and baseline vital signs were taken and recorded: pulse rate was 183bpm, regular and of good volume, peripheral oxygen saturation (SpO_2) was 98% in room air. He was pre oxygenated for 3 minutes using Jackson-Rees circuit at 7L/min. Induction of anaesthesia was by inhalational halothane in 100% O_2 via Jackson-Rees circuit. Intravenous access was achieved using a 20 gauge intravenous cannula on his left hand and trachea was immediately intubated at first laryngoscopy attempt at an adequate depth of anaesthesia using size 3.5mm internal diameter non cuffed portex endotracheal tube (ETT). The tube was strapped at 20cm mark and packing was placed around it after correct placement was confirmed. The ETT was connected to the anaesthetic machine via the Jackson Rees breathing system and oxygen flow rate of 7L/min with 0.5-2% halothane was used for maintenance with intermittent positive pressure ventilation. Intravenous fluid 4.3% dextrose in 0.18% saline was connected to the cannula and administered at 10ml/Kg/Hr in the first hour and 5ml/Kg/Hr subsequently. The patient was carefully positioned for the surgery with hyperextension of the neck and lateral rotation to the left. Care was taken to avoid inadvertent extubation. He was afterwards ventilated to normocarbia and vital signs closely monitored. Analgesia was achieved with intravenous paracetamol 120mg and intravenous pentazocine 4mg.

The intraoperative finding was a right huge cystic cavity with areas of multiloculated which infiltrated to the platysma, sternocleidomastoid muscle, the floor of the mouth, carotid artery, internal jugular vein and ipsilateral parotid gland. There were multiple lymph node enlargements and compressed and deviated trachea. The cyst was completely excised and incision edges closed with a drain left in the cavity. The cyst consisted of a mixture of serous, mucinous, altered blood and fresh blood. Estimated blood loss was 60ml and he was transfused with 160mls of fresh whole blood.

At the end of surgery, his oropharynx was suctioned, halothane was discontinued, and the patient resumed spontaneous breathing and maintained good saturation. He was transferred to the ICU with ETT in-situ when he was cardiorespiratory stable, breathing spontaneously with O_2 entrained into the tube. The ETT was blocked with secretions after 12 hours and it was replaced. The patient, however, developed respiratory distress (respiratory rate = 48 cycles per minute, dyspneic) and was mechanically ventilated using synchronized intermittent mandatory ventilation (SIMV) mode at set rate of 30 breaths per minute, tidal volume of 65ml and positive end-expiratory pressure (PEEP) of 5cmH_2O. He was eventually weaned off mechanical ventilation after 3 days of ICU admission. He received mild sedation with intravenous midazolam 0.5mg hourly. Postoperative analgesia was managed with IV paracetamol 160mg 6 hourly and intramuscular diclofenac 8mg 12 hourly. He was transfused with a second aliquot of blood (120ml). He also received intravenous dexamethasone 0.5mg 12 hourly and intravenous ranitidine 6mg 8 hourly. He was extubated when stable and subsequently transferred to the paediatric ward on the 5th day.

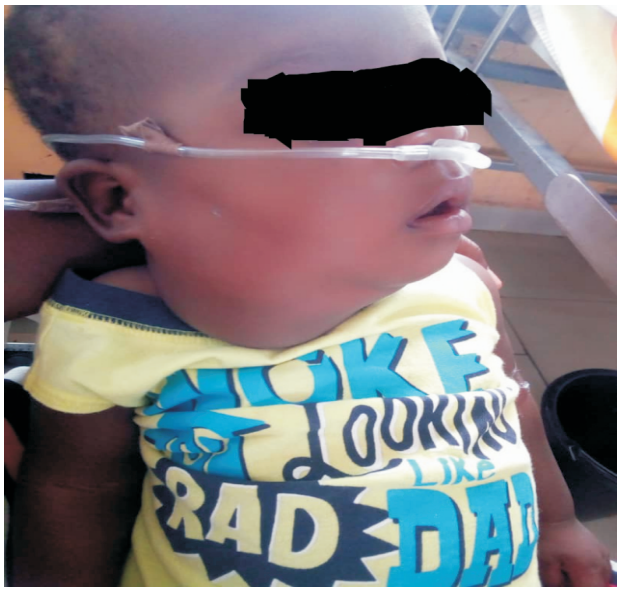

Figure 1 shows the mass on presentation.

DISCUSSION

Cystic hygroma is a congenital cystic tumour caused by a malformation of lymphatic tissue due to failed involution of saccules, obstruction between lymphatics and the venous system, and the sequestered lymphatic rests that penetrate and canalize through fascial planes to contiguous structures. It is also known as cystic lymphangioma, firstly described by Redenbacker in 1828.[1]

Incidence is between 1: 6000 - 16,000 live births, but there is no gender or racial preponderance. It can be diagnosed in-utero, as early as 10 weeks gestational age, using ultrasonography, especially, transvaginally. It can also be diagnosed in the neonates, infants or even among the adults. Ninety per cent of the cases are diagnosed within 2 years of age. In approximately 75% of cases, it is seen in the craniocervical regions, with a 2:1 predilection for the left side. Other areas it can include axilla, mediastinum, groin and retroperitoneum. This patient presented with a neck mass. A strong association has been documented with

Turner's, Noonan, Down's, Edward, and Patau syndromes, [1,2,4,5] though, this patient did not have any of these syndromes.

Surgical excision of cystic hygroma is usually done on elective bases unless it is complicated with severe airway compromise due to sudden enlargement, which may be caused by infection or traumatic bleeding into the mass[2,6,7] as seen in our patient. The case under review was done as an emergency as a result of airway obstruction on presentation. The diagnosis was made at 4months old; however, the excision was not done because the parents did not provide consent for surgery.

It can cause upper or lower airway obstruction or even superior vena cava obstruction. The index patient presented more with supraglottic obstructive symptoms (inspiratory stridor),[4] however, his chest radiograph demonstrated tracheal compression and deviation but no intrathoracic extension. Cervical cystic hygroma usually presents with airway challenges to the anaesthetist, [5] and the anaesthetist should prepare for difficult airway management including difficult airway cart and availability of a surgeon to perform an emergency tracheostomy, should endotracheal intubation fail. Sharma and colleagues[7] reported a case where the surgeon was asked to aspirate the mass before the glottis could be visualized and trachea intubated. Emergency airway management tray and equipment were made available for this patient and in addition, an ENT surgeon was also on standby in the operating suite in case of inability to intubate the trachea.

Awake fibreoptic intubation is the method of choice; however, this may not be possible in infants due to lack of cooperation, and intubation may be more traumatic in them. Inhalational induction is a viable option since it maintains spontaneous ventilation in view of the potential difficult airway. Inhalational halothane was used for induction in this case. Some anaesthetists use suxamethonium and even long-acting muscle relaxants to aid intubation, especially when the patient could be mask ventilated.[5] Heavy sedation should be avoided during premedication, as this may worsen the airway obstruction. The sedative was not used in this patient so as not to worsen the obstructive symptoms.

Adequate preparation should be made for fluid resuscitation and maintenance in view of high fluid shift and blood loss, especially when the mass is huge and infiltrative. Intravenous fluid and blood should be made available.

Effort should be made to prevent hypothermia. In huge and infiltrative mass, the incision is usually wide, which allows heat loss from the patient. The fluids (infusion and blood) to be administered should be warmed.

Positioning is an important concern to the anaesthetist. Usually, the patient is placed in a supine position with hyperextension of the neck and lateral rotation of the neck to the contralateral side. This should be borne in mind to avoid accidental tracheal extubation of the patient. Caution was taken while placing our patient in position in order not to inadvertently dislodge the ETT.

Post-operative airway obstruction is common. This may be caused by haematoma formation, chronically distorted tongue or reactionary oedema. Drains are usually left in place to prevent this. The surgeon left a drain in the cavity. When the tongue is chronically distorted, or there is hypoglossal injury and obstruction by the tongue is anticipated, stitches are used to stabilize it at the dorsum of the tongue. Post-operative airway oedema should be anticipated, especially in extensive surgeries involving the mouth and airway. The patients would benefit from close monitoring in the ICU or high dependency unit (HDU) within the first few days to allow for resolution of the oedema. The tube should be left in-situ. The patient would also benefit from doses of steroid such as dexamethasone, and nebulized adrenaline.

The patient under review was not extubated after the surgery but was sent to ICU for close observation and at the same time, managed with steroid. When he developed respiratory distress, he was mechanically ventilated using SIMV mode for three days before weaning off and finally extubated. This demonstrates the anticipation and preparedness to intervene when the need arises. Leaving the endotracheal tube in situ in the postoperative period facilitated ventilatory support when the patient developed respiratory insufficiency.

CONCLUSION

Airway problems in patients with cystic hygroma constitute a great challenge to the anaesthetist. The mass may distort the anatomy of the airway, causing difficulty in visualizing the glottis and endotracheal intubation. Preparedness is key. The anaesthetist must make provision for difficult airway management. The decision on when to extubate the patient is usually not easy; however, it is safer to leave ETT in-situ for a couple of days to allow oedema to regress.

REFERENCES

1. Acevedo JL, Shah RK, Neville HL, Poole MD. *Lymphatic malformation (cystic hygroma)* [online], Jan. 28, 2019 [Accessed 4th October 2019]. Available from: https://emedicine.medscape.com/article/994055-overview.
2. Kim H, Kim HS, Oh JT, Lee JR. Anaesthetic management for a neonate with giant cystic hygroma involved upper airway-A case report. *Korean Journal of Anesthesiology*. 2011; 60(3): 209.
3. Abdur-Rahman LO, Awolaran O, Nasir AA, Bamigbola KT, Abdulraheem NT, Oyinloye AO, Adeniran JO. Efficacy of bleomycin for non-operative treatment of cervical lymphangioma in University of Ilorin Teaching Hospital, Nigeria. *Journal of Medicine in the Tropics*. 2017; 19(2): 93.
4. Esmaeili MR, Razavi SS, Harofteh HR, Tabatabaii SM, Hosseini HA, Sheikhi MA. Cystic hygroma: Anesthetic considerations and review. *Journal of research in medical sciences: the official journal of Isfahan University of Medical Sciences*. 2009; 14(3): 191.
5. Prasad G, Toshniwal G. Cystic hygroma and anaesthetic implication. *Indian journal of anaesthesia*. 2011; 55(1): 89-90.
6. Lauro HV. *Congenital cysts of the neck (thyroglossal duct cysts including lingual, branchial cleft cyst)* [online], Feb. 15, 2019 [Accessed 4th October 2019]. Available from: https://www.clinicalpainadvisor.com/home/decision-support-in-medicine/anesthesiology/congenital-cysts-of-neck-thyroglossal-duct-cysts-including-lingual-brachial-cleft-cysts.
7. Sharma S, Aminuldin AG, Azlan W. Cystic hygroma: anaesthetic considerations and review. *Singapore medical journal*. 1994; 35(5): 529-31.