

Anaesthetic management of superior vena cava syndrome in a 14-year-old undergoing mediastinal mass excision: a case report

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Abstract

Background: Superior vena cava syndrome occurs when there is obstruction of blood flow through the superior vena cava to the right atrium and is mostly seen in patients with mediastinal masses. It can be a medical emergency associated with respiratory and cardiovascular collapse and requires immediate evaluation and treatment.

Aim: This is a case report of the approach to anaesthetic and intensive care of a 14-year-old scheduled for elective sternotomy for an obstructive mediastinal mass with typical presentations of superior vena cava syndrome.

Case Report: A 14-year-old male patient was diagnosed with features suggestive of superior vena cava syndrome following presentation at the children emergency ward of the University of Port Harcourt Teaching Hospital, Nigeria and after initial management of acute asthma on account of frequent respiratory distress. Following detailed preoperative assessment and preparation, meticulous anaesthetic care and monitoring contributed to a safe outcome despite challenging perioperative cardiorespiratory events typical of patients with such syndrome. The respiratory and cardiovascular variables later returned to normal values following sternotomy and debulking of the mediastinal mass and recovery was uneventful in the Intensive care unit from where he was finally discharged to the ward after 48hrs.

Conclusion: Diagnosis of superior vena cava syndrome can be masked by other common causes of airway obstruction. These patients pose a high anaesthetic risk perioperatively. Management includes surgical approach while specialised anaesthetic care is crucial to a safe outcome.

Keywords: Superior vena cava syndrome, sternotomy, specialised anaesthetic care

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INTRODUCTION

Superior vena cava syndrome (SVCS) is a condition that is associated with obstruction of blood flow through the superior vena cava (SVC) that drains blood from the upper part of the body into the right atrium. This results in oedema and distended veins in the upper part of the body, dyspnoea, orthopnoea etc. It occurs mostly in patients with mediastinal mass enlargement which may be malignant or non-malignant within the thorax, and can

therefore be a medical emergency that requires immediate evaluation and treatment.¹⁻³

William Hunter⁴ first described the syndrome in 1757 in a patient who was diagnosed with syphilitic aortic aneurysm. SVCS is rare in all ages and incidence values in Nigeria are not known, but it is estimated at 1 in 650 to 1 in 3100 patients in the literature; and in the USA about 15,000 cases have been reported yearly.⁵

Tuberculous mediastinitis has also been implicated in the aetiology,⁶ but most cases are

linked to malignancies or metastasis in the thorax⁷ with incomplete SVC obstruction secondary to extrinsic pressure, unlike complete SVC obstruction which is the result of intravascular thrombosis in combination with extrinsic pressure. Such thrombosis may result from intravascular arterial devices and pacemaker wires.^{8,9} The SVC being a thin-walled low pressure vessel in the mediastinum is vulnerable to both extrinsic and intrinsic pressure that leads to blockage of blood flow from the upper body parts and resultant upper extremity oedema, neck vein distension, etc

In adults, malignant causes of SVCS occur mostly in males because of the high incidence of lung cancer in them, but benign causes have no gender predilection.

In the paediatric age, causes may be Non-Hodgkins, or Hodgkins lymphoma, leukaemia, thymoma or infections such as tuberculosis.¹⁰

Conservative treatment may bring some relief, but with decreased cardiac output and cerebral or upper airway oedema, emergency treatment with diuretics or corticosteroids is indicated. Other modalities of management include radio or chemotherapy for some tumors,⁷ thrombolytics or anticoagulants and catheter removal (if possible) when thrombus forms around a central venous catheter, and endovascular treatment options.⁸

Surgical management poses a challenge to the Anaesthetist due to life threatening complications such as cardiovascular collapse; which results from reduced venous return as a result of obstructed flow into the SVC at induction when patient is placed in the supine position. Cerebral oedema with increased intracranial pressure constitute peculiar challenges to the neurological status. Venous access is preferable at the lower limbs due to difficult flow via the upper extremities and presence of a thrombus creates a risk for pulmonary embolism. Complete airway obstruction, pulmonary and laryngeal oedema from venous congestion can be associated with severe hypoxia and difficult intubation during anaesthesia.^{1,2} When all these complications are not well managed, the outcome can be very grave, especially in a

low- and middle-income country where specialized anaesthesia workforce and equipment for monitoring and airway maintenance can be a challenge. Although, there may be need for individualised management, safe practices must be upheld.

We report the successful management of a 14-year-old male in a resource-constrained environment who had been undergoing treatment for repeated bouts of airway obstruction, but was eventually diagnosed with superior vena cava syndrome (SVCS) and booked for an elective mediastinal mass resection.

CASE REPORT

A 14-year-old male student who was 52kg in weight presented to the children's emergency ward of the University of Port Harcourt Teaching Hospital with 3 weeks history of cough and difficulty in breathing of one day duration. The cough was non-paroxysmal, associated with difficult and noisy breathing, productive of whitish sputum and was temporarily relieved by drugs such as antibiotics and bronchodilators. There was no voice change, but there was orthopnoea and daily activities were limited by the difficulty in breathing. There was no previous history of childhood asthma, allergy to drugs or particulate matter, or a positive family history of difficulty in breathing.

Examination of the patient revealed a facial swelling with distended veins on the forehead and over the upper chest. General examination showed him seated upright, dyspnoeic with a respiratory rate of 32cycles/minute and on supplemental oxygen (3l/min) by nasal prongs. There was no fever, pallor, dehydration, central cyanosis, finger clubbing or peripheral oedema. Airway assessment revealed a supple neck with mobile temporo-mandibular joint and an inter incisor distance of >6cm, while the Mallampati assessment for ease of intubation was Grade I. Chest examination showed bilateral chest movement, no swelling or tenderness, percussion notes were resonant, there was markedly reduced air entry with rhonchi in all lung fields. Examination of the cardiovascular system showed a pulse rate of 110 beats/min (regular and of good volume),

blood pressure of 120/80 mmHg, slightly elevated JVP and normal heart sounds I and II.

A chest X-Ray finding that included widening of the mediastinum with a well marginated ovoid mass protruding unilaterally to the right side, and a CT scan that showed a superior mediastinal mass with some narrowing of the SVC at the level of the azygous vein suggested the mass was a thymoma. Other investigations including packed cell volume (34%), serum electrolytes, urea, creatinine and the electrocardiogram results were within normal limits, and an American Society of Anesthesiologists' (ASA) physical status score of IV was assigned.

General anaesthesia (GA) with muscle relaxation, tracheal intubation, and controlled ventilation was planned. Both written and informed consent were obtained and patient was placed on 6hrs preoperative fast for solids and 2hrs to clear fluids. Premedication was omitted, a request was made for 2 units of grouped and cross matched blood and patient was counseled for GA and the likely perioperative course including intensive care unit (ICU) admission for elective post operative ventilation. A multidisciplinary team session that included the Surgical, Anaesthetic, Nursing and Haematologic teams was held preoperatively to discuss envisaged perioperative problems and management plan.

On the day of surgery, following safety checks on the anaesthetic/suction machine, monitors, airway maintenance devices, and the difficult airway tray was placed on standby including plans for emergency tracheostomy, the patient was transported in a propped-up position into the theatre and subsequently positioned on the operating table with monitors (for noninvasive blood pressure, peripheral oxygen saturation, temperature and ECG) attached. Base line vital signs were BP - 125/75mmHg, SpO₂ - 97%, T^oC - 36.8 and ECG - sinus rhythm. Invasive monitoring could not be performed as transducers were not available. Intravenous access was secured on both arms with 16G canulae and warm fluid (0.9% normal saline) administered, but a reduction in flow was observed and an additional access was secured on the right lower limb. Preoxygenation was

commenced with 100% oxygen at a flow of 4L/min for about 5 minutes and IV atropine 0.4mg was administered just prior to induction. Anaesthesia was induced with propofol (100mg) and intubation facilitated with suxamethonium (75mg) using a size 6mm I.D (internal diameter) cuffed endotracheal tube connected to a Mapleson A circuit and equality of air entry confirmed bilaterally. The end tidal capnography (EtCO₂) monitor was also attached and bladder catheterisation instituted to monitor the intraoperative urine output.

Following intubation and before administering the non-depolarising muscle relaxant, there was a sudden increase in intrathoracic pressure, with poor chest compliance and increased air way pressure from the poor breathing (reservoir) bag compliance. The peripheral oxygen saturation (SpO₂) dropped below 90%. Circuits and connections were quickly checked for kinking/disconnections; however auscultation of the chest revealed a marked decrease in air entry bilaterally. Oxygen flow rate was therefore increased to 8-9L/min, inspiratory-expiratory ratio of 3:1 was commenced and respiratory rate was maintained at 16cycles/minute. This improved the SpO₂ to about 95%. Fluid replacement and drug injection could only be achieved via the right lower extremity as there was reverse flow in the upper limb accesses due to increased intra thoracic and central venous pressure.. The BP was also markedly elevated to a range of 140/110 - 180/120mmHg. Anaesthesia was maintained with 100% of oxygen at 3-9l/min and isoflurane at 1-1.5%. Muscle relaxation and manual ventilation with a closed breathing circuit was achieved with 30mg atracurium and multimodal analgesia was employed with IV injections of 20mg pentazocine, 75µg fentanyl and 750mg paracetamol. During sternotomy, manual ventilation was temporarily stopped to prevent damage to inflated lungs from the sternotomy blade. Subsequently, there was an immediate decrease in intra thoracic pressure causing a noticeable reduction in airway pressure with better bag compliance, improved SpO₂ of >95% and lowered BP readings of 120/75 - 130/80mmHg. Also, following debulking of

the tumour, intravenous fluid flow was fully restored on the upper limb and all monitored parameters remained stable, there were no more rhonchi and air entry became adequate bilaterally. The estimated blood loss was 1.2L, patient was transfused with 1L of crossmatched blood and total urine output was about 300mls. At the end of surgery which lasted four and half hours, isoflurane was discontinued and residual neuromuscular block was reversed using 2mg neostigmine and 1mg atropine to obtund its muscarinic effects. Oxygen was continued for another 10min and pharyngolaryngeal suctioning ensured secretions were adequately cleared. The patient maintained strong respiratory efforts with adequate vital capacity breaths, was extubated awake with an oxygen saturation of 96-98% on 100% O₂, and was transferred to the ICU for close monitoring and continued post-operative care. In the ICU, he was nursed in a slight head-up position with supplemental oxygen at 3L/min via nasal prongs, and vital signs were closely monitored continuously, then every 15mins with recorded values within normal range. Pain management continued with IV administration of 20mg pentazocine 8hrly, 750mg paracetamol 6hrly and rectal diclofenac 50mg daily. Temperature maintenance was ensured with warm blankets and breathing exercises were commenced by the second post operative day. He was subsequently transferred to the paediatric ward for continued care after 48hours, and was discharged home on the 20th day post operatively, after an uneventful stay.

DISCUSSION

It is known that no gender predilection occurs with benign causes of SVCS.¹⁰ This index report is on a male gender with thymoma who had a typical presentation of an easily missed early stage SVCS where partial obstruction may be asymptomatic or with minor symptoms and signs. However, with illness progression, the presentation was more classical with a worsening experience on bending forward or lying down.

Anaesthetic management of patients with SVCS must be preceded with a thorough preoperative evaluation which includes a

careful airway assessment. Due to the likelihood of a difficult airway, sedative premedicants are to be avoided and antisialogues administered to dry up secretions. Patient should be transported in a propped-up position (30-45^o) to encourage good venous drainage from the upper half of the body. All these were observed in our patient as documented in previous studies.^{10,11}

Venous access is better secured on the lower extremity to obviate problems related to complete SVC obstruction like pulmonary congestion, increased “one arm –brain circulation” time and a consequent prolonged induction time from reduced time to drug effector site, with the potential risk of over dose of induction agents (and all other drugs in general). Wide bore cannulae (18 or 16G), to manage substantial haemorrhage associated with major intra thoracic surgery is appropriate.¹⁰⁻¹² In our patient however, venous access was initially secured on the upper limbs due to difficulty with alternate access. This could have contributed to the worsening of the SVC obstruction. Kamari et al¹³ have noted a similar finding in their management of a large tumor in the superior mediastinum, where placement of venous access in the upper extremity was attributed as a likely cause of worsening intraoperative SVC obstruction. However, in the present report, the upper limb venous access subsequently became functional and the elevated BP normalised, following sternotomy and tumour resection.

General anaesthesia for SVCS can be associated with increased morbidity and mortality. This is partly caused by partial to total airway obstruction and inability to ventilate the patient. There is also increased risk of difficult intubation as a result of laryngeal edema and engorged veins. Severe haemodynamic compromise is worsened by IPPV leading to increased intra thoracic pressure and decreased venous return.¹¹⁻¹³ Therefore, GA with muscle relaxation should be avoided and maintenance of spontaneous ventilation favoured where feasible.¹⁴ In the present case, GA was conducted being a paediatric, and in order to achieve good

muscle relaxation and airway control. A few studies have employed the use of local anaesthesia, but this was in adults who only had lymph node biopsies or interventional radiological studies.^{11,12} Although difficult intubation was not encountered following induction and muscle relaxation, there was an increase in intra thoracic pressure with difficulty in manual ventilation as the airway pressure was markedly high. However, this was short-lived.

Blood loss may increase from increased venous pressure resulting into hypovolaemia and hypothermia. Blood was therefore replaced and adequate thermal care ensured by warming all fluids and observing a suitable ambient temperature. Renal compromise from hypovolaemia can also be a risk, but adequate fluid replacement and hourly urine output of not less than 0.5-1ml/kg/hr was ensured.

Thereafter, as with most non-complex SVCS's, the increased airway and blood pressures became normalized till discharge. Ideally, postoperative care should be in a paediatric intensive care unit (PICU) for this age category (or a high dependency unit),^{14,15} the study centre however has no PICU and patient was monitored in a general ICU to ensure close monitoring. After an uneventful stay, he was safely discharged to the ward.

CONCLUSION

The anaesthetic management of a patient with SVCS can be very challenging as it is not commonly encountered in clinical practice, Heightened vigilance, careful interpretation of imaging and coordinated multi-disciplinary care are part of indices of favourable outcomes in settings with limited resources as demonstrated in this report.

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Conflict of Interest:

There are no conflicts of interest.

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