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January 2007

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Mohammad Irfan Akhtar Aga Khan University, mohammad.irfan@aku.edu

Mohammad Hamid Aga Khan University, mohammad.hamid@aku.edu

Muneer Amanullah Aga Khan University, muneer.amanullah@aku.edu

Mubashar Khan *Aga Khan University*

Shahabuddin Aga Khan University

See next page for additional authors

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Recommended Citation

Akhtar, M., Hamid, M., Amanullah, M., Khan, M., Shahabuddin, ., Khan, M. (2007). Mid aortic syndrome correction: anaesthetic considerations and management. *Journal of Pakistan Medical Association*, 57(11), 563-565. **Available at:** https://ecommons.aku.edu/pakistan_fhs_mc_anaesth/88

Authors

Mohammad Irfan Akhtar, Mohammad Hamid, Muneer Amanullah, Mubashar Khan, Shahabuddin, and Mansoor Ahmed Khan

Case Report

Mid aortic syndrome correction: Anaesthetic considerations and management

Mohammad Irfan Akhtar, Mohammad Hamid, Muneer Amanullah, Mubashar Khan, Shahabuddin, Mansoor Ahmed Khan Department of Anaesthesia, The Aga Khan University, Karachi.

Abstract

A twelve years old female patient presented with headache off and on and disparity between upper and lower limb pressures. On the basis of history, physical examination and magnetic resonance angiography, the patient was diagnosed as mid aortic syndrome and planned for elective aorto aortic bypass graft surgery through left thoracotomy.

Double lumen endotracheal tube was placed for lung isolation and episodes of hypertension during proximal and distal cross clamp were controlled with sodium nitropruside infusion (SNP). Extra pleural catheter was put in at the end of surgery before chest wall closure for postoperative pain control. Both upper and lower limb pressures were monitored in the cardiac intensive care unit (CICU) and over a period of twenty four hours, gradient gradually reduced to 10 mm Hg.

Introduction

Congenital coarctation of the thoracic aorta at the ligamentum arteriosum or in the aortic arch is well recognized, but a much less common variety of aortic coarctation is located in the distal thoracic aorta, abdominal aorta or both and is called "middle aortic syndrome" or "mid-aortic dysplastic syndrome".¹ A high incidence of both visceral and renal artery stenosis is associated with this condition. Untreated patients may present with severe renovascular hypertension, myocardial infarction, heart failure, intracranial haemorrhage and aortic rupture. Most of the patients if left untreated, would die by 40 years of age. Surgery is the treatment of choice with excellent results although angioplasty has been used with variable success in the last two decades.²

Case Report

A twelve years old female patient, weighing 43 kg, presented in the clinic with history of headache off and on, high blood pressure (BP) in the upper limb and disparity of pulses between upper and lower limbs. Her blood pressure in the upper limb and lower limb was 150/90 and 70/50 mmHg respectively. On the basis of history, clinical examination and magnetic resonance angiography (MRA), she was diagnosed as a case of mid aortic syndrome. Computed tomography (Figure 1) and MRA showed focal

segmental narrowing in mid descending aorta opposite T7 and T8, with good collaterals for splanchnic, spinal cord and lower limb vessels. Trans-thoracic echocardiogram showed a long stenotic segment of descending thoracic aorta with a gradient of 120 mmHg and an incidental finding of supra-valvular aortic stenosis with a 40 mmHg gradient. Her blood pressure was initially controlled with captopril, lasoride and propranolol. She was planned for elective aorto aortic bypass graft surgery through left thoracotomy.

She was admitted in the hospital two days before the surgery to monitor and control her blood pressure. At that time hydralazine was also added for BP control. Patient was premedicated with midazolam 5mg orally one hour before surgery.

In the operating room noninvasive blood pressure, pulse oximeter and electrocardiogram leads were applied. Two 16 guage peripheral intravenous lines were placed. Patient was preoxygenated and induced with fentanyl 100



Figure I. CT-Angiogram demonstrating long segment of narrowed mid thoracic aorta.



Figure 2. Proximal anastomosis of the vascular graft bypassing the stenosed segment in mid thoracic aorta

microgram, etomidate 10 mg and atracurium 30 mg. She was intubated with double lumen tube (28 French) to facilitate surgery. Arterial lines were inserted in right radial and left femoral artery and Central line was placed in right internal juglar vein. Bispectral index (BIS) was applied to monitor the depth of anaesthesia and was kept between 40 and 60. Nasopharyngeal temperature was used for continuous core body temperature monitoring. Patient was put in right lateral decubitus position, taking care of pressure points and neurovascular compression. One lung ventilation started and ventilator set on pressure control of 20, respiratory rate of 15, inspiratory and expiratory ratio of 1:2 and fraction of inspired oxygen (Fio2) 0.5. Episodes of hypertension during proximal and distal cross clamp were controlled with Sodium Nitropruside (SNP) at rate of 0.5 - 3 microgram /kg/min. Anaesthesia was maintained with propofol 2-4 mg /kg/hour and isoflorane 1-2%. Fentanyl was titrated to the total dose of seven microgram/kg. Perfusion distal to cross clamp was monitored by femoral arterial line and urine output. Intraoperative blood gases showed pH 7.35, PCO2 42, PO2 109, HCO3 22, Na 135, k 3.7 and Hb 7.7 gm/dl. Aorto aortic graft was put in successfully as shown in the Figure 2. Patient remained stable throughout the procedure. Extra pleural catheter was put in at the end of surgery before chest wall closure and continuous infusion of Bupivacaine 0.25% started at the rate of 8-10 ml/hour.

Total blood loss was 550 ml and two units of packed red blood cells transfused during the procedure. Arterial blood gas at the end surgery showed, Ph 7.36, PCO2 41 mmHg, PO2 84 mmHg, HCO3 22 mEq/L, Base excess -2.6, Hb11 gm/dl, Hct 32, K+ 3.97 mEq/L and Na+ 135 mEq/L. Proximal cross clamp time was 50 minutes and distal cross clamp time was 60 minutes. Urine output during clamping was 40 ml and mean lower body pressure was 30-40 mmHg.

Patient was extubated and shifted to cardiac intensive care unit (CICU) with supplemental oxygen and routine monitoring. In the CICU, blood pressure surges were controlled with Sodium Nitropruside (SNP) infusion. Extra pleural catheter and intravenous analgesics were used for pain control. Both upper and lower limb pressures were monitored in CICU and over a period of twenty four hours the gradient reduced to 10 mmHg.

Discussion

Middle aortic syndrome is a rare entity which is characterized by stenosis in the descending thoracic aorta and comprises of 0.5% to 2.0% of all the Coarctations.³ The condition is usually associated with some form of vasculitis especially Takayasu's but it may occur in isolation.³ The condition is usually diagnosed in young adults but may present in childhood as a challenging problem⁴ of managing hypertension. Stenting⁵ and surgery⁶ has been performed to treat this condition.

Anaesthetic plan should include perioperative blood pressure control, maintenance of distal perfusion during cross clamp, spinal cord protection, and anticipation of excessive blood loss and its appropriate replacement, application of one lung ventilation and postoperative pain control. In the present case, preoperatively the blood pressure was controlled with combination of ACE inhibitor, Beta blocker, loop diuretic and a direct acting arterial dilator. Inraoperatively, upper body hypoperfusion was controlled with SNP while lower body hypoperfusion before cross clamping. Fortunately, our patient had adequate collaterals which prevented lower body hypoperfusion. Postoperative pain was addressed with 0.25% Buivacaine infusion through extra pleural catheter.

Recognized complications of surgery are haemorrhage, paradoxical post-operative hypertension and paraplegia. The incidence of paraplegia is about 3 to 9% and prolonged cross lamp time is an important factor in the development of this complication. Long term results after surgery are satisfactory in the absence of arteritis however persistent hypertension⁷ and reno-vascular disease appear to affect life expectancy.

Conclusion

Appropriate anaesthetic management of mid aortic syndrome correction requires preoperative planning and anticipation of intraoperative adverse outcome. Management should include perioperative blood pressure control, maintenance of distal perfusion during cross clamp, spinal cord protection, anticipation of excessive blood loss and its appropriate replacement, application of one lung ventilation and postoperative pain control.

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