Considerations for Takayasu Arteritis Undergoing General Anesthesia in a Resource-Limited Setting: Case Report and Literature Review

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Introduction

Takayasu arteritis, also called “aortic arch syndrome”, “young female arteritis”, and “pulseless disease”, is a rare chronic progressive large-vessel vasculitis of unknown etiology that affects the aorta and its major branches [1]. The clinical presentation of Takayasu arteritis patients vary greatly, which often makes definitive diagnosis difficult and delayed. The disease predominantly affects Asian women during the second or third decade of life and is associated with considerable morbidity and premature mortality attributed to a delay in diagnosis [2,3]. The anesthetic management of patients with Takayasu arteritis is well described with most of the literature involving patients who had undergone surgery under neuraxial anesthesia [4-10]. Furthermore, reports have emphasized the importance of utilizing intraoperative monitoring devices such as invasive arterial blood pressure and cerebral perfusion monitoring devices [10-12], which are unavailable in our institution when this case was performed. We therefore present our experience of a patient with Takayasu arteritis who had undergone general anesthesia in a resource-limited setting.

Case Report

A 23-year-old man (height, 163 cm; weight, 37 kg; BMI, 13.9) was scheduled for endoscopy-guided turbinoplasty for septal deviation with turbinoseptal contact of the right middle turbinate and posterior area of the nasal septum. His history of present illness started nine months prior to surgery when the patient consulted an otorhinolaryngologist due to frequent episodes of nasal congestion accompanied by pain between the eyes, headache and dizziness that were not relieved by oral medication. After undergoing panendoscopy, the doctor attributed his symptoms to a deviated nasal septum and advised turbinoplasty. His past medical history only consisted of an uncomplicated hemorrhoidectomy done in 2011. He was an otherwise healthy man with unremarkable personal and social history and no family history of anesthetic problems or other inheritable conditions.

On physical examination by the attending physician, it was noted that his pulse was feeble on the left arm precluding blood pressure measurement using a conventional sphygmomanometer and stethoscope. On the other hand, pulses were audible on his right arm and his blood pressure (BP) was noted to be 100/60 mmHg. Laboratory findings were unremarkable. Twelve-lead ECG revealed left ventricular hypertrophy and 2-D echocardiography showed mild regurgitation and mild...
mitral valve prolapse with an ejection fraction of 69%. The patient was referred to a cardiologist who observed similar physical examination findings in addition to a decreased left carotid pulse.

A computed tomographic angiography was done which revealed an occlusion of the left common carotid artery and left subclavian artery. There is a short segment stenosis on the proximal celiac artery, as well as double renal artery supply of the right kidney, an ectatic left renal artery, and a left-sided inferior vena cava in the abdominal region. The aorta was found to be normal with no evidence of stenosis, aneurysm, or dissection. The brachiocephalic artery, right common carotid artery and right subclavian artery were patent. The patient was subsequently diagnosed with Takayasu arteritis and he was started on a combination therapy consisting of prednisolone, methotrexate and folic acid.

Before surgery, the patient was premedicated with oral midazolam. Once in the operating theater, automated noninvasive BP monitors were placed on the patient’s right upper and lower limbs. Electrocardiogram (5-lead) and finger pulse oximeter were also placed. Baseline BP of 100/60 mmHg on the right upper limb and 120/70 mmHg on the right lower limb were recorded before the induction of anesthesia.

After proper preoxygenation, general anesthesia was induced using intravenous midazolam 1 mg, fentanyl 50 mcg, lidocaine 20 mg, and propofol 100 mg. This was followed by administration of intravenous atracurium 25 mg and oral intubation was performed using a McCoy blade laryngoscope. Anesthesia was maintained with sevoflurane 1.5% with oxygen and intermittent boluses of phenylephrine were given as needed to maintain mean arterial pressure within 20% of the preoperative values. He was also given an intravenous dose of acetaminophen 1 g and tramadol 50 mg as preventive analgesia. The patient remained hemodynamically stable throughout the 2-hour procedure, sustaining only minimal blood loss. After recovery of neuromuscular blockade and adequate spontaneous breathing, the patient was given intravenous lidocaine 80 mg prior to deep extubation. The entire postoperative course was uneventful and the patient was discharged on the fourth postoperative day.

Discussion

Takayasu arteritis is a rare chronic intractable disease of unknown etiology characterized by vasculitis involving mainly the aorta and its major branches. It is the arterial inflammation that causes stenosis, occlusions or aneurysmal dilatations that predispose the patient to symptomatic end-organ ischemia [13-15]. The clinical presentation of patients with Takayasu arteritis, therefore, varies greatly depending on the extent of vascular involvement and the degree of disease progression. Considerable morbidity and premature mortality are attributed to a delay in diagnosis; thus, a high index of clinical suspicion is emphasized [2,14,16].

As for epidemiology, Takayasu arteritis is most commonly seen in Japan, South East Asia, and India [17]. In the Philippines, the reported combined number of cases of Takayasu arteritis at the University of the Philippines-Philippine General Hospital and Philippine Heart Center is 61 during the years 2006-2015 [18]. It is now recognized worldwide that the disease can affect both sexes and may have an onset of > 40 years of age [19,20]. Moreover, a recent study reported a larger proportion of Takayasu arteritis patients who are male and with an elderly onset [14]. However, the disease may also present in childhood [21,22].

Classification of the disease can be radiographic based on angiographic findings or clinical based on symptoms or complications [23,24]. Ishikawa [23] described the four most important complications as secondary hypertension, retinopathy, aneurysm formation, and aortic regurgitation. Hypertension is the most common complication observed in a Filipino multicenter cohort [18].

Cited descriptions on the anesthetic management of patients with Takayasu arteritis were mostly of parturient women who have undergone successful neuraxial anesthesia [8,9,25-28]. Irrespective of the choice of anesthetic technique, pre-anesthetic assessment must initially take into account the extent of affected arteries and degree of organ involvement, as intraoperative anesthetic concerns are often related to severe uncontrolled hypertension, stenosis or aneurysms of blood vessels that may affect regional circulation predisposing to end-organ dysfunction, and difficulties in monitoring the blood pressure [11,29]. A history of dizziness or syncope on extension of the head may be a clinical feature suggestive of carotid involvement that compromises cerebral perfusion; hence, is an important consideration when laryngoscopy is contemplated [7,11].

Preoperatively, standard ASA monitoring is adequate for most of these patients [7,11,26,28,30]. The use of intraoperative cerebral perfusion monitoring devices has been found to be of clinical value to detect ischemia among patients with cerebral artery stenosis [10,31-33]. This is important as postoperative cerebral infarction can occur in this patient population [34]. Hypertensive parturient women are especially vulnerable and the use of regional cerebral oxygen saturation guided cerebral monitoring during delivery has been described [35]. Nonetheless, in patients with undocumented cerebrovascular defects, there have been several reports of uncomplicated neurological outcomes in which general anesthesia was used without these devices [7,11,12,21,30,33,36].

The key concept of perioperative anesthetic management is to maintain adequate blood flow to vital organs by avoiding significant blood pressure fluctuations especially severe hypertension. Both noninvasive and invasive blood pressure monitoring have been described in literature [11,12,21,26,27,32-34,36-39]. Blood pressure is best measured from the extremity least affected by the disease process. Monitoring of upper and lower extremities is advocated only when there is a discrepancy of more than 20 mmHg [7,25]. Meikli and Milne, however, suggested that monitoring of both upper and lower blood pressure should be considered to have some measure of overall perfusion.
[40]. Other methods of noninvasive blood pressure monitoring that have been reported include the use of Doppler blood flow signals and visual estimation of systolic blood pressure using plethysmographic waveform by pulse oximetry [29,41]. Intra-arterial monitoring of blood pressure may be necessary if extreme and rapid blood pressure fluctuations are anticipated from the surgery or if no blood pressure recordings are attainable from any extremity [12,21,33,36]. Another study recommended monitoring of beat-to-beat variations in blood pressure as a proactive measure to correct mean arterial pressure intraoperatively [27]. It is further advised that in inserting the catheter, the tip should be placed proximal to the area of arteritis, as there is a difference in blood pressure proximal and distal to the area of arteritis. Central system blood pressure was reported to be 100 to 120 mmHg over and above those obtained in the upper limbs peripherally [42].

Because the patient has an occlusion on the left main artery of the neck, we used a McCoy blade laryngoscope to intubate the patient to reduce the head extension angle that may predispose the patient to cerebral ischemia. The McCoy laryngoscope, introduced in 1993, is a modification of the Macintosh laryngoscope. It is a levering laryngoscope that has a hinged tip that is operated by a level mechanism on the back of the handle which permits epiglottic elevation [43]. In hospitals where fiberoptic bronchoscopes or video laryngoscopes are not available, the McCoy laryngoscope is a useful alternative in visualizing the airway in patients with limited neck extension [44]. In our case, the attending anesthesiologist happened to own a McCoy laryngoscope. In cases where a McCoy laryngoscope is unavailable, intubation can still be achieved using a Macintosh laryngoscope [33]. Although limited evidence suggests that head extension during intubation in patients with cervical disorders may cause secondary injuries [45], a prompt smooth intubation is advised as it can also help prevent prolonged unwanted hemodynamic stress responses that can be detrimental to high-risk patients with Takayasu arteritis.

Postoperatively, means to avoid hypertension associated with extubation and acute pain should be taken. Moreover, postoperative pain relief is essential to prevent undesirable stress-related complications. The decision to transfer the patient to an ICU and the length of postoperative anesthesia care unit (PACU) stay or close monitoring should be tailored according to the severity of the disease and to the risk of developing further complications.

In conclusion, patients with Takayasu arteritis vary greatly in terms of clinical presentation; thus, meticulous preoperative evaluation, intraoperative planning, and postoperative management are necessary. We have presented here a case of a young male with an incidental diagnosis of Takayasu arteritis who had undergone uneventful surgery under general anesthesia aided by a basic understanding of the uncommon disease.

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Conflict of Interest

None declared.

References

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