Anesthetic Management of Klippel-Trenaunay-Weber Syndrome (KTWS)

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Abstract

Klippel-Trenaunay Weber Syndrome (KTWS) is a rare congenital neuroectodermal disorder characterized by a triad of varicosities, cutaneous capillary abnormalities, and soft tissue hypertrophy in affected limbs. KTWS is often associated with arteriovenous malformations. The syndrome has been linked to genetic mutations in angiogenic factors that have not yet been well defined. A 15-year-old patient with KTWS that underwent repair of a left radicular arteriovenous fistula originating at T11-L2 and the anesthetic challenges involved with her care is described in this case study. Her medical history was significant for capillary malformations, multiple arteriovenous fistulas of the left lower extremity and significant soft tissue hypertrophy in the affected extremity. The ability to provide safe anesthesia care for a patient with KTWS requires the practitioner to gain a full understanding of the syndrome and how it differs from similar syndromes. In developing a course for anesthesia administration in this unique patient population, it is critical for the anesthesia provider to become familiar with the regions of the body affected by KTWS, particularly in the lumbar spine, oral cavity, and airway, since this knowledge will ultimately guide the anesthetic plan.

Keywords: Klippel-Trenaunay Syndrome (KTS), Klippel-Trenaunay-Weber Syndrome (KTWS), congenital vascular nevus, capillary hemangiomas, cutaneous capillary malformation
**Introduction**

Klippel-Trenaunay Weber Syndrome (KTWS) is a rare congenital neuroectodermal disorder characterized by a triad of varicosities, cutaneous capillary abnormalities, soft tissue hypertrophy in affected limbs and is often associated with arteriovenous malformations (AVM).\(^1,^2\) Anesthetic management of patients with KTWS is challenging due the high incidence of bleeding associated with regional anesthesia, which generally eliminates this option due to safety concerns. Anesthetic management is further complicated by the possibility of poor control of hemodynamic responses to laryngoscopy, which could lead to a rupture of an intracranial AVM or severe bleeding from the oral cavity if vascular malformation is present.

**Case Report**

A 15-year old, Hispanic female presented for repair of a left radicular arteriovenous fistula originating at T11-L2. Her medical history was significant for capillary malformations, multiple arteriovenous fistulas of the left lower extremity and significant soft tissue hypertrophy in the affected extremity. The patient’s only prior anesthetic or surgical history included an open repair of an arteriovenous fistula involving the tibial artery of the lower left extremity which was done under spinal anesthesia. Information obtained from the patient's mother included the patient had no known medical drug allergies and was not taking any prescription or over-the-counter medications. The physical examination was unremarkable, including an airway evaluation. Vital signs preoperatively: Wt: 67.4 kg, Ht: 171 cm, Temp 36.4 C, HR 87 beats/min, BP 144/78 mmHg, and RR of 20 breaths/min. The patient's Mallampati Classification (MP) was rated as a MP 1 with a thyromental distance of greater than three finger breadths. The patient was categorized as an American Society of Anesthesiology class III. Her preoperative laboratory
values were; hemoglobin 13.7 g/dL, hematocrit 39.2%, platelet count 189 x 1000/mm³, blood glucose 98 mg/dL, PT 11.2 seconds, PTT 30.6 seconds, and an INR of 1.1.

The anesthetic plan consisted of administration of total intravenous anesthesia (TIVA) using propofol and remifentanil in order to facilitate somatosensory evoked potential (SSEP) monitoring during the operative phase. Numerous failed attempts at intravenous (IV) access due to a plethora of venous malformations led to an unacceptable patient anxiety level, resulting in a clinical decision to defer IV access and adding mask induction to the anesthesia plan of care. Following pre-oxygenation, a mask induction using sevoflorane with a 70% nitrous oxide (NO) 30% oxygen (O₂) mixture was performed.

Two 18-gauge intravenous catheters were placed in both forearms, followed by an IV induction of fentanyl 100 mcg, lidocaine 60 mg, etomidate 14 mg, and succinylcholine 100 mg. Intubation with placement of a 7.0 mm endotracheal tube (ETT), with a Glidescope® (Verathon Inc., Bothell, WA) was completed without complication. Immediately following intubation a 20-gauge right radial arterial line was placed and TIVA using propofol and remifentanil was initiated. Anesthesia was maintained during the remainder of the seven hour operative case without complications or loss of SSEPs. As soon as surgical closure began, one minimum alveolar concentration (MAC) of sevoflurane was initiated in combination with the administration of 1 mg of diluadid while the TIVA was weaned in preparation for extubation. Dexamethasone 10 mg IV and ondansetron 4 mg IV were both administered prophylactically for prevention of postoperative nausea and vomiting (PONV). Upon conclusion of the procedure the patient was extubated without complication. The patient was stable, alert, oriented to person,
place and time. The patient was then transferred to the Neurocritical-Care (NCC) unit for observation.

**Discussion**

KTWS is a syndrome marked by congenital malformations ranging from minimally significant to life threatening. The exact etiology and pathogenesis of KTWS are unknown; however, multiple theories exist. Most cases are sporadic, although a few cases in recent literature suggest an autosomal dominant pattern of inheritance.³

The capillary malformations are present at birth and progress proportionally with physical growth. These malformations typically localize to extremity, with soft tissue and osseous hypertrophy of the affected region as well as development of multiple, microscopic; fast-flow arteriovenous shunts. Although usually associated with an extremity, KTWS malformations can occur anywhere throughout the body. Ultrasonography, magnetic resonance imaging (MRI) and magnetic resonance arteriography (MRA) are often used to differentiate between Klippel-Trenaunay Syndrome (KTS) and KTWS.⁴

In 1900, noted French physicians Klippel and Trenaunay first described a syndrome in two patients presenting with a port-wine stain and varicosities of an extremity associated with bony and soft tissue hypertrophy. In 1907, Parkes and Weber, unaware of Klippel and Trenaunay’s report, described a patient with the same presentation as well as an arteriovenous malformation of the affected extremity.¹ ² Today, conflicting opinion exists in the literature regarding whether to separately designate the original triad as Klippel-Trenaunay syndrome and the triad with the addition of arteriovenous malformation (AVM) as Parkes Weber syndrome. Making the
distinction is probably wise given the increased morbidity associated with arteriovenous malformations.\textsuperscript{1} A small but extremely important subset of affected individuals also harbors AVMs on the spine.\textsuperscript{5}

The ability to provide safe anesthesia care for a patient with KTWS requires the practitioner to gain a full understanding of the syndrome and how it differs from similar syndromes. In developing a course for anesthesia administration in this unique patient population, it is critical for the anesthesia provider to become familiar with the regions of the body affected by KTWS, particularly in the lumbar spine, oral cavity, and airway, since this knowledge will ultimately guide the anesthetic plan. Regional anesthesia is generally avoided in these patients due to the possibility of AVMs of the lumbar spine.\textsuperscript{6}

During the initial consultation between the anesthesia and surgical teams regarding this 15-year old female’s operative course, the inquiry of airway evaluation via MRI or ultrasound was questioned. Chart review revealed a MRI had been obtained four years prior and was clear of airway malformation or defects. Patients with KTWS should always be considered to have potentially difficult airways due to frequent presence of soft tissue hypertrophy, upper airway angiomas, and facial anomalies.\textsuperscript{7}

Given the anatomical concerns associated with KTWS and the time since the last MRI had been obtained, it was determined a Glidescope® (Verathon Inc., Bothell, WA) would be used for intubation and ETT placement to decrease the possibility of bleeding. Utilizing this method
would also provide an opportunity for more than one individual to assess and evaluate the airway.

Another primary concern for the anesthesia team was to ensure the patient remained normotensive during induction and throughout the case. Fluctuations in blood pressure leading to a hypertensive state could potentially cause rupture of multiple intracranial and peripheral arteriovenous shunts and capillary malformations. Complications of KTWS can include lymphedema, and internal bleeding from vascular abnormalities and fistulas, all of which can be exacerbated by elevated blood pressure.7

Maintaining a normotensive state was a priority given the patient had to be positioned prone during the operation. Prone positioning during anesthesia is associated with predictable changes in cardio- pulmonary physiology. In the prone posture, pressure on the abdomen compresses the inferior vena cava and femoral veins, diverting blood from the distal parts of the body into perivertebral venous plexuses.8 A critical part of the anesthetic plan was preparation for unforeseen vascular complications such as hypertensive and hypotensive states. Nitroprusside sodium, dopamine hydrochloride and phenylephrine intravenous drips were pre-mixed before the case began.

The final issue to be addressed as part of the anesthesia plan was the possibility of a deep vein thrombosis (DVT) becoming dislodged during the operation. Due to the venous abnormalities associated with KTWS and the presence of soft tissue hypertrophy in the patient’s left lower extremity, the likelihood of suffering from a DVT was very high. Recurrent DVTs and
pulmonary emboli (PE) have been described in multiple studies and are quite common. The literature is unclear regarding recommendations in patients that have a high probability of experiencing DVTs during the intraoperative or postoperative phase. Patients with KTWS are not candidates for anticoagulation therapy given the high risk of bleeding with traditional therapies. Further studies are needed to evaluate the possibility of using low-molecular weight heparin in this population.

In conclusion, it has been demonstrated in this case, as in others that with appropriate planning regarding hemodynamics, blood loss, and airway control, complex and lengthy surgeries can be performed safely in patients with KTWS.
References


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