Case Report

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Airway and Anaesthetic Managment in Patients with Klippel-Treanunay Syndrome

Sushmitha S[1], Kiran N[2]

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Abstract

Introduction: Klippel–Trenaunay syndrome is a rare sporadic condition described in terms of a clinical triad, Atypical lateral varicosity, Port wine stain (capillary malformation), Bone and soft tissue hypertrophy or hypotrophy.

Case Report: We present a 20-year-old male, Non Vysya patient presented with history of pain in left ear and yellowish white sero-purulent discharge in the left ear since 10 days. On local examination there was discoloration over the left side of the face in the maxillary region, bifid tongue, swelling over the inner aspect of the left side of the mouth, the face was elongated with hypertrophy of the soft tissue of the left side of the face.

Patient was diagnosed to have Left ear Chronic Otitis Media with postauricular abscess and was posted for Incision and drainage, canaloplasty, meatoplasty and excision of intraoral lesion. Anaesthesia was maintained with the help of Propofol, oxygen, nitrous oxide and vecuronium. Patient was fully awake and extubated, also we were able to see vocal cords movement during extubation. Patient was shifted to Post Anaesthesia Care Unit.

Conclusion: Patients with Klippel–Trenaunay syndrome is rare condition needs thorough understanding of hemodynamic changes, proper preoperative assessment with vigilant intraoperative monitoring and management.

Keywords: Klippel-Trenaunay syndrome; Port wine stain; Video Laryngoscopy; Malformations.

Key Message: Klippel-Trenaunay syndrome is a rare syndrome where we can anticipate difficult airway. Hence, vigilant management during pre-operative, intraoperative and post-operative is necessary.

Author's Affiliation: [1]Junior Resident, [2]Professor, Department of Anaesthesiology, Sri Devaraj URS Medical College, Tamaka, Kolar 563101, Karnataka, India.

Corresponding Author: Kiran N., Professor, Department of Anaesthesiology, Sri Devaraj URS Medical College, Tamaka, Kolar 563101, Karnataka, India.

E-mail: dr.nkiran@gmail.com

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INTRODUCTION

Klippel –Trenaunay syndrome is a rare sporadic condition described in terms of a clinical triad,[1] Atypical lateral varicosity[2] Port wine stain (capillary malformation)[3] Bone and soft tissue hypertrophy or hypotrophy. It involves capillary, venous and lymphatic vascular malformations. It is congenital overgrowth disorder discovered by Paul

Trenaunay in 1900. Lung, Gastrointestinal tract, liver, kidney and bladder can also have venous malformations and hemangiomas and could lead to life threatening complications like haemorrhage. Pulmonary thromboembolism is also seen in patient having such syndrome. Finally it can also lead to intracerebral haemorrhage and compression neurological structures in spinal cord.

CASE REPORT

A 20-year-old male, Non Vysya patient presented with history of pain in left ear and yellowish white sero-purulent discharge in the left ear since 10 days. On local examination there was discoloration over the left side of the face in the maxillary region, bifid tongue, swelling over the inner aspect of the left side of the mouth, the face was elongated with hypertrophy of the soft tissue of the left side of the face. Patient had regular pulse (86 beats per min), Blood pressure of 120/70 mmHg and respiratory rate of 14/min. Electrocardiogram showed normal sinus rhythm. MRI brain showed Malignant Otitis Externa. True congenital hemifacial hypertrophy and chronic infarcts in left superior colliculi and vermis of cerebellum. Patient was diagnosed to have Left ear Chronic Otitis Media with postauricular abscess and was posted for Incision and drainage, canaloplasty, meatoplasty and excision of intraoral lesion.

Routine investigations were done and they were well within normal range. Pre-operatively, one wide bore 18G cannula was secured. Patient was given nebulization and nasal packing done with Lignocaine 2% and adrenaline, after shifting the patient to Operating theatre, nasal pack was removed and Lignocaine 4% spray was sprayed.

Continuous ECG, Noninvasive blood pressure, saturation, capnography monitoring was done. Monitors were connected, pre-oxygenation was done for 3 minutes. Premedication was done using Injection Glycopyrrolate 0.2mg + Injection Midazolam 2mg + Injection Fentanyl 100mcg was given, oral pack with pads were done to ensure proper maximum ventilation, Propofol 100mg+20mg+20mg was given and ventilation with bag and mask was checked, then Injection Succinylcholine 100mg was given and intubation done. Video laryngoscope was used. With some alteration in head posture and video laryngoscope with bougie, we were able to intubate with Eight mm Endotracheal tube orally, Bilateral air entry was equal. In a separate I.V cannula, small doses of Dexmedetomidine Infusion were given to maintain hemodynamics. Anaesthesia was maintained with the help of Propofol, oxygen, nitrous oxide and vecuronium. Patient was fully awake and extubated, also we were able to see vocal cords movement during extubation. Patient was shifted to Post Anaesthesia Care Unit.

DISCUSSION

The trio of venous malformations or varicose veins, cutaneous capillary abnormalities, and bony or soft tissue hypertrophy characterise Klippel Trenaunay syndrome, a rare congenital abnormality with a frequency of 1 out of 27,500 live births.[4,5] varicose veins or venous malformations, and bony or soft tissue hyperplasia of an extremity. It is one of many heterogeneous disorders known as overgrowth syndromes that are characterized by either generalized or localized somatic overgrowth. Overgrowth syndromes each have unique clinical, behavioral, and genetic features, but some of these features overlap, causing diagnostic difficulty. Cutaneous manifestations, however, can be key to distinguishing the various syndromes. We present a patient with an unusual variant of KTS consisting of right upper extremity hyperplasia, lymphedema, and cutaneous and visceral lymphangiomas. We review several closely related syndromes and discuss the differential diagnosis of limb hyperplasia.Vascular anomalies may impact the lung, trunk, gastrointestinal system, bladder, and neurovascular tissues.[2] Lee JH et al., presented with case of massive bleeding in KTS patients for close reduction and internal fixation for femur shaft fracture correction.[1]

The risk of difficult airways (facial abnormalities, upper airway angiomas, and soft-tissue hypertrophy of the airway), the possibility of severe intraoperative bleeding, local intravascular coagulation within the malformation, disseminated intravascular coagulation, consumptive coagulopathy, and thrombocytopenia are the anaesthetic concerns in patients with KTS.[5,6] capillary malformations that may involve neurovascular structures, and bony or soft tissue hypertrophy in affected limbs. Areas such as the trunk, bowel, bladder, and spinal cord may be involved as well. KTS should not be confused with Klippel-Feil syndrome, which involves abnormalities of the cervical vertebrae. Anesthetic management for patients with KTS has only been described in limited case reports that caution about potential airway difficulty but do not report surgical hemorrhage requiring transfusion.

METHODS

We performed an electronic search of the Mayo Clinic medical record database to identify patients who had undergone an anesthetic for surgery related to KTS. Review of medical records was performed for type of surgery, anesthetic technique, airway management and difficulty, medications used, intraoperative fluid administration, transfusion requirements, vascular access used, and postoperative complications.

RESULTS

Eighty-two unique patients were identified who underwent 134 general anesthetics and 2 lumbar neuraxial anesthetics for surgeries related to KTS. Preoperatively, 27% of patients had a history of recurrent bleeding, 24% recurrent cellulitis, 9% deep vein thrombosis, and 2% pulmonary embolism. The mean age at time of surgery was 21 ± 15 years. The majority of surgical procedures involved laser coagulation or varicose vein sclerotherapy or stripping. All of the 74 direct laryngoscopies and tracheal intubations were performed on the first attempt without difficulty. Mask ventilation was possible in all 131 patients for whom this was attempted, with only 1 requiring an oral airway. Documented estimated blood loss ranged from 20 to 18,000mL, with a mean of 740 ± 2739mL. Use of a tourniquet did not obviate the possibility of substantial blood loss. The only significant postoperative complication involved a calf hematoma after vein stripping and avulsion that required return to the operating room for evacuation.

CONCLUSIONS

Patients with KTS have multiple associated comorbidities relevant to perioperative management. In contrast to previous reports, difficulty with airway management was not encountered. Surgery related to severe KTS may be associated with massive hemorrhage despite tourniquet use, and the anesthesiologist should anticipate the need for appropriate fluid resuscitation. intravascular coagulation takes place inside the malformation in KTS, and coagulation factors are being depleted in the distal flow.[7] Disseminated intravascular coagulation and KMS (consumptive coagulopathy and thrombocytopenia) are also often linked to KTS[3] As a result, the anesthesiologist needs to exercise caution when it comes to having enough intravenous access, a sufficient reserve of blood products, and proper intraoperative monitoring. Thirdly, there are several risk factors associated with central neuraxial blocking, including haemangiomas, spinal arteriovenous malformations, coagulation problems, and venous dilatation that may result in epidural hemorrhatomas.[5,8]soft tissue and bony hypertrophy and varicose veins. In addition, venous malformations or hemangiomas may also be found in the lung, gastrointestinal tract, liver, kidney and bladder, which may result in recurrent hemor-rhage and compromise the affected organs. Involvement of the cerebrum, cerebellum or spinal cord may lead to intracerebral bleeding and compression of neurological structures. Venous thrombosis and pulmonary thromboembolism are both common and may cause pulmonary hypertension and right ventricle failure. In some patients, in addition to flat hemangiomas, cavernous hemangiomas can be seen with a rapid growth rate in the first year of the patient's lifespan, producing high-output congestive heart failure and consumptive coagulopathy (Kasabach-Merritt syndrome).

Ultrasound guidance can be used to rule out vascular abnormalities of the jugular/subclavian veins if central venous pressure monitoring is to be performed. Given lower limb thrombophlebitis associated with venous abnormalities, it is preferable to avoid femoral cannulation. Because brain haemangiomas increase the risk of intraoperative bleeding, precautions against blood pressure spikes must be implemented.[8] The anesthesiologist can guarantee a positive outcome for KTS patients having surgery by using the previously outlined measures.

Conflict of Interest: None

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