Case Report

Anaesthesia management in a child with Klippel-Trenaunav Syndrome posted for examination of both eyes under general anaesthesia: A case report

Sirisha Senthil[1], *Harshitha Kadava*[2], *Hari Shanker Charan*[3], *Raja Narsing Rao*[3].

[1]Department of Cataract and Glaucoma, LVEPI, Hyderabad, [2]Department of Glaucoma, LVEPI, Hyderabad, [3]Department of Anaesthesia, LVEPI, Hyderabad

Abstract

Klippel-Trenaunay syndrome (KTS) is a very rare congenital vascular anomaly. It is characterized by the presence of capillary malformation, venous malformation as well as limb overgrowth, generally affecting one extremity. Although clinical characteristics of KTS are well known, the epidemiology and pathophysiology still remain to be defined. Awareness of this disorder is important for anesthesiologist for managing potential complications. Here, we report a case of five-months old male baby posted for examination of both the eyes and Transscleral cyclophotocoagulation (TSCPC) under general anaesthesia.

Key words:

general anaesthesia, Klippel-Trenaunay syndrome, examination of eyes, arterial venous malformation

Address for correspondence:

Dr. Raja Narsing Rao
Consultant Anaesthesiologist
Department of Anaesthesia, LVEPI
Hyderabad
Email: rajahadigal@gmail.com

Article History

Received: 15th November 2021
Revision: 20th November 2021
Accepted: 21st December 2021
Published: 17th January 2022

Introduction

Over a hundred years ago, French physicians Klippel and Trénaunay described for the first time a rare congenital disorder named Klippel–Trénaunay syndrome (KTS)[1] with a very low incidence of about 1:100,000.[2] KTS is a capillary–lymphatic–venous malformation associated with soft-tissue and skeletal hypertrophy and it is clinically recognized by a triad of capillary malformations (port wine stain), atypical venous malformations and bony and/or soft-tissue hypertrophy; presence of any two of these features will confirm the diagnosis.[3]

The syndrome is usually diagnosed at birth, but it can be found in older children and adults if not diagnosed in time. Extremities, particularly the lower extremities, are affected by vascular abnormalities.[4] Most cases of KTS are sporadic, but little has been published about family members suggesting an inherited disorder.[2,5]

Case description

We report a case of five-months old baby who had with a whitish opacity over the cornea of the left eye (LE). Opacity was noted since 3 days after birth and was gradually increasing in size.

How to cite this article: Sirisha Senthil, Harshitha Kadava, Hari Shanker Charan, Raja Narsing Rao. Anaesthesia management in a child with Klippel-Trenaunav Syndrome posted for examination of both eyes under general anaesthesia: A case report Indian J Ophthal Anaesth 2022;2(1):35-8

Pinkish discoloration of the skin was seen on different parts of the body associated with swelling of middle, fourth and little toes of both the feet, Figure 1 and 2. It was full-term caesarean delivered baby with birth weight of 2.4 kg and cried soon after birth.

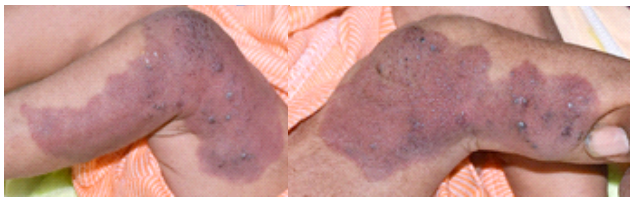

Figure 1. Capillary Hemangiomas involving lateral aspect of both legs

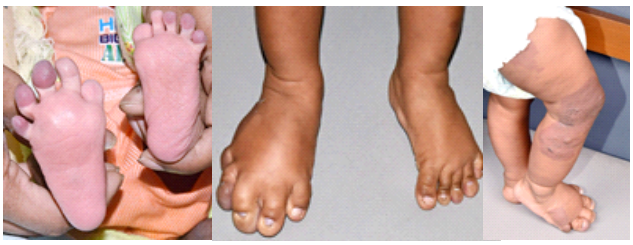

Figure 2. Hemangiomas involving middle, 4th and little toes of both feet with soft tissue overgrowth

The baby underwent a thorough pre-anaesthetic evaulation by a pediatrician. On examination, both cardiovascular and respiratory system was normal. Routing blood and urine investigations were also found to be within the normal limits. Universal protocol, 4-2-1 for nil per oral was followed. On the day of surgery, the baby was shifted into the operation theater and standard monitoring ECG, peripheral oxygen saturation and non-invasive arterial blood pressure was attached. Anaesthesia was induced with 50% O_2:50% N_2O + 6% Sevoflurane. Intravenous line was established. Airway was secured with appropriate sized AMBU LMA and the baby was on spontaneous assisted mode of ventilation. The vitals were stable throughout the procedure. After the procedure was over, gentle suctioning was done and the baby was extubated in left lateral position.

The baby was kept under observation for one hour and handed over to the parents after uneventful adequate recovery.

Discussion

KTS has no predilection for gender, race or geographical area and occurs sporadically.[2,6] Although the etiology of KTS is still unknown, damage to the sympathetic nervous system resulting in dilatation and persistence of microscopic arteriovenous anastomoses *in utero* is hypothesized to be the leading cause.[7] KTS has been shown to belong to a spectrum of segmental overgrowth diseases caused by mutations in the PIK3CA gene, which differentiates it from Parkes–Weber syndrome (PWS), which is caused by mutations of the RASA1.

Alteration of tight balance between angiogenesis and vasculogenesis occurs. Most patients with KTS will present with the classic triad[8] but some clinical variations can be seen with age (childhood and adulthood),[9] however, these are far less common. Few numbers or absence of lymphatic channels are leading causes of lymphedema that can be documented using ultrasound (with or without doppler) and magnetic resonance imaging (MRI).[10] Extremities are mostly affected, most often unilaterally (85%), sometimes bilaterally (12.5%), and only rarely crossed-bilaterally (2.5%).[12] The case presented in this report had right lower limbs affected.Various other limb anomalies including camptodactyly, syndactyly, clinodactyly, and congenital hip dislocation have been reported in association with KTS.[10]

Spectrum of PIK3CA Related Overgrowth Spectrum (PROS)[8]

- KTS
- CLOVES- Congenital Lipomatous Overgrowth, Vascular malformations, Epidermal nevi, Scoliosis

- MCAP- Megalencephaly Capillary Malformation
- HHML- Hemi Hyperplasia- Multiple Lipomatosis
- FAO- Fibro-adipose Overgrowth

The presence of arteriovenous fistula in PWS is the only difference with KTS, and both syndromes are generally confirmed with Doppler ultrasound and magnetic resonance angiography.[10,11]

These patients may require orthopaedic surgeries to correct limb discrepancies, debulking surgeries and amputations; interventions like sclerotherapy and laser therapy for vascular malformations, vascular surgeries like surgical stripping and endoscopic ligation of perforating veins, and surgical resection of the bowel in case of gastrointestinal haemorrhage.

Preoperative cardiology evaluation must be ensured as patients may have venous thrombophlebitis (50%) and pulmonary thromboembolism (22%) which may lead to pulmonary hypertension and right ventricular failure.[12] Arteriovenous malformations can produce high output congestive heart failure. Preoperatively, deep vein thrombosis prophylaxis must be considered. Enough packed RBC should be ready before the operation for appropriate fluid resuscitation in case of haemorrhage. In certain cases, preoperative embolisation can be carried out to reduce intraoperative bleeding. This will need close collaboration with interventional radiologists.

Reports concluded that general anaesthesia is safe for patients with KTS. Difficult intubation must be anticipated as patients may have facial anomalies, upper airway angiomas and soft tissue hypertrophy in the airway. Loss of auto-regulation in the abnormal vessels predisposes to hemorrhage, especially in the presence of hypertension that may occur intra-operatively. Measures to obtund the hemodynamic responses to direct laryngoscopy and tracheal intubation, noxious surgical stimuli, and extubation should be performed. It is important that the patient is normotensive during induction and throughout the operation. Fluctuation of blood pressure might lead to a hypertensive state that could potentially cause rupture of multiple intracranial and peripheral arteriovenous shunts, aneurysms and capillary malformations. Other complications such as internal bleeding from vascular abnormalities and fistulas, might be exacerbated by elevated blood pressure. Maintaining a normotensive state is important if the patient is positioned prone during the surgical operation. Prone positioning is associated with predictable changes in cardiopulmonary 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 peri-vertebral venous plexuses. Essential part of the anaesthetic plan is the preparation for unexpected vascular complication such as hypertensive and hypotensive states. Nitroprusside sodium, dopamine hydrochloride and phenylephrine infusions should be kept ready before surgery. Excessive venous pulsation can result in inaccurately low pulse oximetry reading if the probe is placed on an affected limb. Avoidance of coughing, straining, retching, and vomiting is important to prevent rupture of the abnormal vessels. Coughing and bucking should be avoided during extubation. Central neuraxial blockade should be consider with caution due to risk of epidural hematomas in the presence of haemangiomas, spinal arteriovenous malformations and neurovascular malformation in the surrounding structure

of the spine; tendency for coagulation disorders, and venous dilation. If neuraxial blockade is planned, it is mandatory to do preoperative CT/MRI to rule out vascular malformations in the CNS and ensure absence of cutaneous lesions overlying site of needle insertion. Preoperative coagulation profile must be done in these patients.

Postoperative monitoring should be individualized depending on the surgical procedure done, the preoperative status, and the intraoperative complications.

Currently, there is no definitive treatment of KTS approved; however, a multidisciplinary management should be focused on reducing symptoms and complications associated to the disease.

Conclusion

We presented the case of an infant with a rare congenital vascular disorder type KTS who presented with glaucoma vascularmalformationon the left eye, as well as numerous port wine stains. This case serves as review of clinical features and etio-pathology of KTS and also highlights the importance of a multidisciplinary management team and follow-up, which can help to avoid the occurrence of complications that have an impact on the prognosis of the patient.

References

1. Klippel M, Trenaunay P. Du naevus variquex osteohypertrophique. Arch Gen Med. 1900;3:641–72.
2. LacerdaLda S, Alves UD, Zanier JF, Machado DC, Camilo GB, Lopes AJ. Differential diagnoses of overgrowth syndromes: the most important clinical and radiological disease manifestations. Radiol Res Pract. 2014;2014:947451
3. Jacob AG, Driscoll DJ, Shaughnessy WJ, Stanson AW, Clay RP, Gloviczki P. Klippel–Trenaunay syndrome: spectrum and management. Mayo Clin Proc. 1998;73(1):28–36.
4. Garzon MC, Huang JT, Enjolras O, Frieden IJ. Vascular malformations. Part II: associated syndromes. J Am AcadDermatol. 2007;56(4):541–64.
5. Aelvoet GE, Jorens PG, Roelen LM. Genetic aspects of the Klippel–Trenaunaysyndrome.Br J Dermatol. 1992;126(6):603–7.
6. Ivanitskaya O, Andreeva E, Odegova N. Prenatal diagnosis of Klippel–Trenaunay syndrome: series of four cases and review of the literature. Ultrasound. 2020;28(2):91–102.
7. Gloviczki P, Driscoll DJ. Klippel–Trenaunay syndrome: current management. Phlebology. 2007;22(6):291–8.
8. Vahidnezhad H, Youssefian L, Uitto J. Klippel–Trenaunay syndrome belongs to the PIK3CA-related overgrowth spectrum (PROS). ExDermatol. 2016;25(1):17–9.
9. Eerola I, Boon LM, Mulliken JB, et al. Capillary malformation-arteriovenous malformation, a new clinical and genetic disorder caused by RASA1 mutations. Am J Hum Genet. 2003;73(6):1240–9.
10. Redondo P, Bastarrika G, Aguado L, Martinez-Cuesta A, Sierra A, Cabrera J, Alonso-Burgos A. Foot or hand malformations related to deep venous system anomalies of the lower limb in Klippel–Trenaunay syndrome. J Am AcadDermatol. 2009;61(4):621–8.
11. Jacob AG, Driscoll DJ, Shaughnessy WJ, Stanson AW, Clay RP, Gloviczki P. Klippel–Trénaunay syndrome: spectrum and management. Mayo Clin Proc. 1998;73(1):28–36.
12. Baba A, Yamazoe S, Okuyama Y, Shimizu K, Kobashi Y, Nozawa Y, Munetomo Y, Mogami T.A rare presentation of Klippel–Trenaunay syndrome with bilateral lower limbs. J Surg Case Rep. 2017;2017(2):rjx024.