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Anaesthestic Management of a Patient with Klippel Trenaunay Syndrome for RF Ablation of Varicose Veins

Dr. Kishanth kumar S[1], Dr. Sanjana K.V. L[2], Dr. Lakshmi[3]

[1]Post graduate, Department of Anesthesiology and pain medicine, Saveetha Medical College and Hospital
[2]Assistant professor , Department of Anesthesiology and pain medicine, Saveetha Medical College and Hospital
[3]Head of the department, Department of Anesthesiology and pain medicine, Saveetha Medical College and Hospital .

Abstract: Klippel Trenaunay Syndrome (KTS) presents unique challenges for anaesthetic management, particularly during procedures such as radiofrequency ablation (RFA) of varicose veins. This abstract outlines the considerations and strategies employed in the anaesthetic management of a patient with KTS undergoing RFA. Key factors such as vascular anomalies, coagulopathy, and potential airway difficulties are discussed. The utilization of preoperative assessment, meticulous planning, and close intraoperative monitoring is emphasized to ensure optimal outcomes and minimize complications. Collaboration between anaesthesiologists, vascular surgeons, and other healthcare professionals is crucial in managing these complex cases effectively. This abstract highlights the importance of a multidisciplinary approach and tailored anaesthetic techniques in providing safe and successful care for patients with KTS undergoing RFA for varicose veins.

1. Introduction:

Klippel-Treanunay syndrome (KTS) is a rare congenital overgrowth disorder associated with vascular anomalies first described by Paul Treanunay in 1900 . It is usually associated with complex vascular malformations such as cutaneous hemangiomas and varicosities in extremities. Such lesions tend to bleed easily because of the fragility of the blood vessels Congenital malformation characterized by the association of soft tissue and bony hypertrophy, venous malformations, lymphatic abnormalities, and cutaneous capillary malformations.
A port-wine stain and venous varicosities in association with bony and soft tissue hypertrophy, radiology (elongation and cortical thickening of affected bones), and arterial and venous evaluation (angiography, venography) are characteristic. Histology shows capillary spread of the papilla dermis adjacent to the lesion, but also in deeper layers of dermis and subcutis. Associated anomalies include ocular anomalies, glaucoma, cerebral aneurysm, spinal cord arteriovenous malformations, gastrointestinal hemorrhage, and severe menorrhagia
Spinal arteriovenous malformations may be associated with the cutaneous haemangiomas in the same dermatomal distribution as the cutaneous lesion. Central neuraxial blockade is fraught with risk due to haemangiomas and spinal arteriovenous malformations, tendency for coagulation disorders, and venous dilation that can cause epidural haematomas. Therefore ,it is traditionally recommended that spinal and epidural anaesthesia is avoided in these patients.

2. Case Report:

A 16 year old boy who is a known case of Klippel trenaunay syndrome came with complaints of varicose veins in B/L lower limbs , Has port-wine stains , disproportionate extremities,Hypertrophy of limbs ,height -174 cm , weight-58kg ,No known co-morbidities ,The procedure lasted a total of 45 mins associated with very minimal blood loss.Intraoperatively patient's vital parameters and hemodynamic status were stable.Patient was shifted to Post Anesthesia Care Unit and continued monitoring.In the post-operative period, evaluation of patient revealed full recovery of motor blockade with power of 5/5 in both lower limbs after 2 hours. Pre op investigations were within normal limits.Patient was planned for B/L radiofrequency Ablation of varicose veins under Spinal anesthesia., Patient was shifted inside the OR, routine monitors connected, 1 18G venflon was secured and pre loaded with IVF to prevent post SAB hypotension, Basal heart rate was 90 bpm, blood pressure was 120/80 mmHg and SpO2 of 98% at room air.The Subarachnoid block was given using 26G spinal needle at L3-L4 space,

and 2 ml 0.5% hyperbaric bupivacaine with 25mcg fentanyl as adjuvant was used under aseptic precautions. Adequate sensory and motor blockade was achieved. B/L LL RF ablation done successfully .

3. Management:

Difficult intubation must be anticipated as patients may have facial anomalies, upper airway angiomas and soft tissue hypertrophy in the airway.
The potential for massive intra-operative haemorrhage must be considered. In KTS, local intravascular coagulation occurs within the malformation, and distal flow has depletion of coagulation factors.
KTS is also often associated with disseminated intravascular coagulation and KMS (consumptive coagulopathy and thrombocytopenia).
The anaesthesiologist must, therefore, be vigilant with sufficient intravenous access, adequate blood product reserve and appropriate monitoring intra-operatively.
Central neuraxial blockade is fraught with risk due to haemangiomas and
spinal arteriovenous malformations, tendency for coagulation disorders, and venous dilation that can cause epidural haematomas.
However, it must be noted that central regional blockade has been carried out safely after ruling out vascular malformations in the central nervous system with computed tomography/magnetic resonance imaging, and ensuring absence of cutaneous lesions overlying the site of needle insertion.

4. Discussion:

Klippel Trenaunay syndrome (KTS) is a vascular malformation syndrome comprising varying involvement of cutaneous capillaries, veins, and lymphatics with hypertrophy of soft tissue and bones of the affected limb. This syndrome is also referred to as capillary-lymphatic-venous malformation (CLVM). This condition was first described in 1900 by two French physicians, Maurice Klippel and Paul Trenaunay. KTS is a clinical diagnosis made by the presence of at least two of the three classic findings of localized cutaneous capillary malformations, venous abnormalities, and limb hypertrophy.
The presence of arteriovenous malformations is now considered a separate entity named as Parkes-Weber syndrome, distinct from KTS Etiology of Klippel Trenaunay syndrome to somatic mutations in the hosphatidylinositol-4-5-bisphosphate 3 kinase, catalytic subunit (PIK3CA) gene. This leads to the activation of phosphatidylinositol-3-kinase (PI3K)/protein kinase and cell overgrowth by dysregulation of the mTORC2 pathway. Mutations occur in the embryological stage of development involving angiogenesis, reflecting findings seen in this condition. Now, KTS is grouped under the umbrella of similar overgrowth syndromes – PIK3CA-related overgrowth spectrum (PROS).

5. Conclusion:

We would like to conclude that, provided the appropriate imaging study was first performed, patients with Klippel-Trenaunay syndrome need not be denied central regional anaesthetic techniques.

6. References:

1. 1.Kumar S, Taneja B, Saxena KN, Kalra N. Anaesthetic management of a neonate with Kasabach-Merritt syndrome. Indian J Anaesth. 2013;57:292–4.
2. Gober-Wilcox JK, Gardner DL, Joste NE, Clericuzio CL, Zlotoff B. Limb hyperplasia: Case report of an unusual variant of Klippel-Trenaunay syndrome and review of the literature. Cutis. 2009;83:255–62.
3. Barbara DW, Wilson JL. Anesthesia for surgery related to Klippel-Trenaunay syndrome: A review of 136 anesthetics. Anesth Analg. 2011;113:98–102.
4. Pereda Marín RM, García Collada JC, Garrote Martínez AI, Miralles Serrano EM, Morales Aguilar JL. Anesthetic management of Klippel-Trénaunay syndrome and attendant gastrointestinal hemorrhage. A case report. Minerva Anestesiol. 2007;73:187–90.