KLIPPEL-TRENAUNAY SYNDROME: A TROUBLESOME CONDITION

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BACKGROUND

Klippel-Trenaunay syndrome (KTS) is a rare congenital disorder characterized by the triad of port-wine stains, varicose veins and bony/soft tissue hypertrophy involving an extremity. Besides the limbs, other areas may be affected, such as the spinal cord and neck/oropharynx, which pose important challenges to the anesthesiologist: **airway management**, **controversy about neuraxial techniques** and **potential blood loss**. Because of its rarity and the importance of the anesthetic management considerations it requires, we report a case at our institution.

CASE REPORT

- 47-year-old women, ASA III
- Past medical history: KTS, pulmonar thromboembolism, anticoagulation with rivaroxaban, hemithyroidectomy and neoadjuvant radiotherapy
- Scheduled for **excision of popliteal sarcoma**
- Preoperative evaluation: no signs of difficult airway, normal echocardiogram and blood analysis. Interruption of rivaroxaban two days before surgery

Fig. 1 – Port-wine stain typical of KTS

Anesthetic management:

- ✓ General anesthesia
- ✓ Endotracheal intubation under direct laryngoscopy without difficulties
- ✓ Hemodynamic stability and pain control was maintained and no significant blood loss was recorded
- ✓ Neuraxial technique was NOT performed → No preoperative imaging studies were done to exclude neurovascular involvement

Fig. 2 – Excision of popliteal sarcoma

DISCUSSION

KTS incidence is low (1:27500)¹ and its anesthetic management is still a matter of controversy. Despite previous reports of **difficult airway management**, a recent study¹ didn't encounter significant airway difficulties, which was also the case of our patient. **Blood loss** is a potential problem, especially because these patients are frequently anticoagulated, but our patient didn't register significant haemorrhage. We didn't performed any **neuraxial technique** because no neuroimaging study was made before surgery. Still, this possibility can be considered in specific conditions, even if it is controversial².

LEARNING POINTS

- **Potential difficult airway** is possible (even if rare)
- **Massive blood loss** should be anticipated
- **Neuraxial techniques** are controversial: may be considered only if vascular malformations were excluded by neuroimaging