ANESTHETIC MANAGEMENT OF THE KLIPPEL-TRENAUNAY WEBER SYNDROME

Dra. Sánchez, M^a Elena; Dra. Gispets, Dolors; Dra. Sánchez-Migallón, Virginia;
Dra. Boliart, Yolanda; Dra. Borràs, Rosa M^a; Dr. Miranda, Alejandro

Hospital Universitario Dexeus (Grupo Quirón-Salud)

Background:

Klippel-Trenaunay-Weber syndrome (KTWS) is a rare congenital neuroectodermal disorder characterized by the triad of **varicose veins, cutaneous capillary abnormalities and soft tissue hypertrophy**. Frequently, it is associated with arteriovenous malformations (AVM). It has been linked to genetic mutations in angiogenic factors that are not yet well defined.

Case Report:

A 12-year-old patient diagnosed with KTWS admitted for scheduled surgery of proximal and distal epiphysiodesis of the left tibia and fibula. A general anesthesia is planned, discarding the neuraxial technique due to the existence of alterations in the lumbar blood vessels.

Once in the operating room, pneumatic stockings are placed on the patient as thromboembolic prophylaxis is done. Surgery is performed under balanced general anesthesia. Induction is performed with Fentanyl 100mcg, Propofol 100mg and Rocuronium 30mg. A first direct laryngoscopy is performed in which no lesion is observed at the level of the oral cavity and an orotracheal intubation is performed without evidence of bleeding.

For anesthetic maintenance, Sevoflurane was used at fCAM 1%, remaining hemodynamically stable during surgery, without needing vasoactive drugs. The surgery proceeds without any incident and the patient is extubated in the operating room.

Conclusions:

KTWS is a **challenge at the anesthetic level** due to the **high incidence of complications**. The development of an adequate and exhaustive plan, familiarizing itself with the affected body regions (lumbar spine, oral cavity and airway), minimizes the risk in these patients.

The **safest anesthetic management** depends on a **deep knowledge of the disease and its implications**.

Bibliografía:

- Renard D, Campello C, Taieb G, et al. Neurologic and vascular abnormalities in Klippel- Trenaunay-Weber syndrome. JAMA neurology. Jan 2013;70(1):127-128.
- Barbara DW et al. Anesthesia for surgery related to Klippel-Trenaunay syndrome: a review of 136 anesthetic. Anesthesia and analgesia. Jul 2011;113(1):98-102.
- Abd-Elseyed AA et al. Anesthetic considerations in a parturient with Klippel- Trenaunay-Weber Syndrome Society of obstetrics anesthesia. 2011:Abstract #109.

CONGENITAL CONECTIVE TISSUE DISORDERS

Collagen synthesis disorders
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Alterations in tissue resistance
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- Vascular malformations and hemangiomas
- Thrombocytopenia and/or anemia
- Valvular diseases
- High-grade heart insufficiency
- Polydactyly or syndactyly

REGIONAL ANESTHESIA

High incidence of bleeding
(presence of vascular malformations or alterations in coagulation)

GENERAL ANESTHESIA

- Possible poor response to hemodynamic changes secondary to laryngoscopy: intracranial AVM rupture or oral cavity bleeding
- Avoid drugs that depress cardiac contractility