orphanesthesia

Anesthesia recommendations for

Kasabach-Meritt syndrome or phenomenon

Disease name: Kasabach-Meritt syndrome or phenomenon

ICD 10: D75.8

OMIM: 141000

Synonyms: Hemangioma-thrombocytopenia syndrome, Kaposiform hemangioendothelioma

Disease summary:

The Kasabach-Merritt phenomenon is the association of a rapidly enlarging vascular lesion anywhere on the body (commonly an extremity but also head and neck, thigh, sacrum, retroperitoneum) with consumptive coagulopathy (low fibrinogen, increased D-dimers) and thrombocytopenia because platelets are trapped into the tumor. Hemolytic microangiopathic anemia is also frequently present. It is exclusively associated with the vascular tumors kaposiform hemangioendothelioma (KHE) and tufted angioma (TA). These are benign vascular tumors that typically present in infancy, and are classified as having intermediate malignant potential as they are locally aggressive but are not known to metastasize. The Kasabach-Meritt phenomenon may affect up to 70% of all patients with KHE and up to 10% of patients with TA [1,2].

KHE and TA are vascular tumors made of irregular nodules with a complex vascular architecture. KHE presents as an enlarging, firm, purpuric cutaneous or soft tissue lesion with poorly defined margins and with spindle-shaped cells (active phase) at histology. TA presents as a series of violaceous macules and papules with a glomerular structure with crescentlike vascular cleft, before the active phase or during the regression phase at histology. Some accompanying lymphangiomatosis can be observed [1,2].

When the Kasabach-Meritt phenomenon starts, KHE or TA rapidly enlarges and becomes tense, purpuric, or ecchymotic, and painful. The swelling may be dramatic.

On the contrary from infantile hemangiomas (common vascular malformation) the endothelial cells lining the lesion are negative for the glucose-transporter-1 (GLUT-1) isoform and for the Lewis Y (LeY) antigen.

Careful surgical excision is performed when feasible but the lesion is often extensive, and a medical treatment is necessary: it includes high-dose steroids, vincristine, α- or β- interferon and platelet aggregation inhibitors such as ticlopidine and/or aspirin [3,4,5,6]. Propranolol is sometimes used with success [5]. Sirolimus with or without steroids is currently often used with excellent results.

Platelet transfusion is best avoided because it generally results in enlarging the lesion and worsening the coagulopathy because of platelet trapping into the tumor [1,2,6,7,8,9].

The lesion usually appears during the first year of life. Mortality varies between 10 and 40%. Death usually occurs from life-threatening hemorrhage, cardiac failure, or invasion of the vascular lesion into local structures [2,6|.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

Translations may not always reflect the most recent updates of the English version

Find more information on the disease, its centers of reference and patient organizations on Orphanet: www.orpha.net

A	**AIRWAY / ANESTHETIC TECHNIQUE**	Thrombocytopenia usual: regional anesthesia may be contraindicated. Risk of difficult mask ventilation and or intubation if the upper airway is involved by the tumor.
B	**BLOOD PRODUCTS (COAGULATION)**	Check hemoglobin, platelets, fibrinogen and D-dimers. High risk of coagulopathy. Blood, fibrinogen and platelets available? Avoid platelets transfusion. Tranexamic acid for prophylaxis and treatment of coagulopathy.
C	**CIRCULATION**	Preoperative echocardiography: signs of heart failure? sequelae of chemotherapy? Check blood potassium if hemolytic anemia.
D	**DRUGS**	NSAIDs to be used with caution. Tranexamic acid ready for use.
E	**EQUIPMENT**	Thromboelastography useful in case of bleeding. Blood warming device.

Typical surgery and procedures

Biopsy or excision of the lesion, placement of central venous access for chemotherapy, embolization of the vessels feeding the tumor, tracheostomy in case of upper airway involvement.

Type of anesthesia

General anesthesia for procedures related to the vascular lesion or for unrelated procedures. Locoregional anesthesia is rarely feasible because of thrombocytopenia. Procedural sedation and analgesia with extreme caution in case of head and neck involvement.

Necessary additional pre-operative testing (beside standard care)

Blood count and coagulation screen, including D-dimers [7,8,9].

CT-scan or MRI to define the extension of the lesion.

Consultation of a specialist in vascular malformations and in pediatric oncology.

Side effects of chemotherapy: electrolytes, renal and liver function tests, echocardiography

Side effects of sirolimus therapy: systemic hypertension, microcytic anemia, hypertriglyceridemia

Particular preparation for airway management

Airway involved by the lesion: possible difficult intubation and/or extubation [7].

Airway not involved: gentle airway management to avoid bleeding (e.g. in case of nasal intubation in the presence of thrombocytopenia) [7].

Particular preparation for transfusion or administration of blood products

Platelet transfusion has to be avoided [7].

Correction of consumption coagulopathy: fibrinogen, tranexamic acid, cryoprecipitate or prothrombin concentrate may need to be administered; recombinant activated Factor VII (rFVII) may be needed in case of bleeding unresponsive to previous treatments: remember fibrinogen level must be normal for rFVII to be effective [7].

Correction of anemia with packed red blood cells [7].

Thromboelastrography may be very useful to correct a coagulopathy [7].

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transportation and mobilization

Not reported.

Interactions of chronic disease and anesthesia medications

The child's comorbidities should be taken into account.

No specific drug interaction described but the side effects of chemotherapy have to be taken into account.

Avoid using NSAIDs in the presence of thrombocytopenia.

Beware of risk of bradycardia or inappropriate hemodynamic response to hypovolemia in case of preoperative propranolol therapy.

Supplemental steroids in case of preoperative corticosteroid therapy.

Anesthetic procedure

All types of anesthesia can be performed. However, in case of planned locoregional or neuraxial anesthesia, be aware of thrombocytopenia.

Particular or additional monitoring

Standard but adapted to the invasiveness of the procedure undertaken.

Preoperative echocardiography in case of heart failure or risk of cardiotoxicity following chemotherapy.

Possible complications

Uncontrollable hemorrhage and hemodynamic collapse.

Rapid enlargement of the lesion.

Airway obstruction if the lesion involves the head and neck.

Heart failure in case of massive arteriovenous shunting across the lesion.

Post-operative care

To be adapted to the invasiveness of the procedure undertaken and the size/location of the lesion.

Disease-related acute problems and effect on anesthesia and recovery

Disease-triggered emergency situations in the Kasabach-Merritt syndrome are:

- acute thrombocytopenia: consumption of platelets in the lesion vs dilutional coagulopathy?
- acute or progressive airway obstruction in case of involvement: brisk tumoral enlargement versus glottic or subglottic edema?
- cardiac dysrythmias: check plasma potassium level, especially in the presence of anemia: acute hemolysis?

Ambulatory anesthesia

Probably unsafe unless the lesion is stable and quiescent (no thrombocytopenia) and the procedure does not involve the tumor; overnight stay to be foreseen in case of doubt.

Obstetrical anesthesia

Not reported.

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