orphanaesthesia

Anaesthesia recommendations for

Kasabach-Merritt syndrome

Disease name: Kasabach-Merritt syndrome

ICD 10: D75.8
OMIM 141000

Synonyms: Haemangioma-thrombopaenia syndrome, kaposiform haemangioendothelioma

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) with consumptive coagulopathy (low fibrinogen, increased D-dimers) and thrombopaenia because platelets are trapped into the tumor.

The lesion is a vascular tumor made of irregular nodules presenting as a kaposiform haemangioendothelioma (spindle-shaped cells) (active phase) or a tufted angioma (glomerular structure with crescentlike vascular cleft, before the active phase or during the regression phase) pattern and with some accompanying lymphangiomatosis at histology.

On the contrary from infantile haemangiomas (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 platelets antiaggregants such as ticlopidine and/or aspirin.

Platelet transfusion is best avoided because it generally results in enlarging the lesion and worsening the coagulopathy because of platelet trapping into the tumor.

The lesion usually appears during the first year of life. Mortality is around 10%.

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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

Typical surgery

Excision of the lesion, placement of central venous access for chemotherapy, tracheostomy in case of upper airway involvement.

Type of anaesthesia

General anaesthesia for procedures related to the vascular lesion or for unrelated procedures. Locoregional anaesthesia rarely feasible because of thrombopaenia. Analgo-sedation 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. CT-scan or MRI to define the extension of the lesion.

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

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

Particular preparation for airway management

Airway involved by the lesion: Possible difficult intubation and/or extubation.

Airway not involved: Gentle airway management to avoid bleeding (e.g. in case of nasal intubation).

Particular preparation for transfusion or administration of blood products

Platelet transfusion has to be avoided.

Correction of consumption coagulopathy: Fibrinogen, tranexamic acid.

Particular preparation for anticoagulation

Not reported.

Particular precautions for positioning, transportation and mobilisation

Not reported.

Interactions of chronic disease and anaesthesia medications

Not reported.

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

Anaesthetic procedure

All types of anaesthesia can be performed.

In case of planned locoregional or neuraxial anaesthesia, be aware of thrombocytopaenia.

Particular or additional monitoring

Standard but adapted to the invasiveness of the procedure undertaken.

Echocardiography in case of heart failure or risk of cardiotoxicity following chemotherapy.

Possible complications

- Uncontrollable haemorrhage
- Enlargement of the lesion
- Airway obstruction if the lesion involves the head and neck
- Heart failure in case of massive arteriovenous shunts 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 anaesthesia and recovery

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

- acute thrombopaenia: 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 oedema?

Ambulatory anaesthesia

Probably unsafe unless the lesion is stable and quiescent (no thrombopaenia); overnight stay to be foreseen in case of doubt.

Obstetrical anaesthesia

No case reported.

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