orphanaesthesia

Anaesthesia recommendations for patients suffering from

Kasabach-Merritt syndrome

Disease name: Kasabach-Merritt syndrome

ICD 10: D75.8

Synonyms: haemangioma-thrombopenia syndrome, Kaposiform haemangioendothelioma

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, D-dimers) and thrombopenia because platelets are trapped into the tumor.

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

On the contrary from infantile haemangiomas (common vascular malformation) the lesion is negative for glucose-transporter-1 (GLUT-1) 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 in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic 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

Locoregional anaesthesia rarely feasible because of thrombocytopaenia

Analgo-sedation with extreme caution in case of head and neck involvement

Necessary additional diagnostic procedures (preoperative)

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, transport or mobilisation

Not reported.

Probable interaction between anaesthetic agents and patient's long term medication

Not reported.

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

Anaesthesiologic procedure

All types of anaesthesia can be performed.

In case of planned Locoregional or neuraxial anaesthesia be aware of thrombocytopenia.

Particular or additional monitoring

Standard but adapted to the invasiveness of the procedure undertaken.

Echocardiography in case of heart failure.

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 shunt across the lesion.

Postoperative care

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

Information about emergency-like situations /Differential diagnostics

caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the diseases, e.g.:

Disease triggered emergency-like situations are not common in Kasabach-Merritt syndrome.

Ambulatory anaesthesia

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

Obstetrical anaesthesia

Not reported.

Literature and internet-links

1. Enjolras O, Wassef M, Mazoyer E et al. Infants with Kasabach-Merritt syndrome do not have “true” hemangiomas. J Pediatr 1997; 130: 631-40.
2. Wassef M, Vanwijck R, Clapuyt P et al. Vascular tumours and malformations, classification, pathology and imaging. Ann Chir Plast Esth 2006; 51: 263-81.
3. Enjolras O, Picard A, Soupre V. Congenital haemangiomas and other rare infantile vascular tomours. Ann Chir Plast Esth 2006; 51: 339-46.

Last date of modification: June 2011

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