orphanaesthesia

Anaesthesia recommendations for patients suffering from

Kikuchi-Fujimoto disease

Disease name: Kikuchi-Fujimoto disease

ICD 10: I88.1 Lymphadenitis cervical, non specified site, chronic or subacute

Synonyms: Histiocytic necrotizing lymphadenitis, Kikuchi disease, Kikuchi-Fujimoto disease, Kikuchi lymphadenitis, lymphadenopathy, KFD

KFD is a rare lymphohistiocytic disorder due to cervical inflammatory lymphadenitis (or axillary region and even other location seldom) with an unknown etiopathogenesis, that is most commonly seen in young Asiatic people (male/female=1:1, despite some data suggest it is more frequent in women). Some HLA class II genes are more frequent in patients with KFD. In particular, the incidence of DPA1*01 and DPB1*0202 alleles is significantly higher in patients with KFD than in healthy control subjects. It is mainly characterized by lymphadenopathy, fever, nocturnal sweats, myalgia, weight loss, and arthralgia, and commonly follows a self-limited course. Less frequently symptoms observed include cutaneous lesions, hepatosplenomegaly, central nervous system impairment and hemophagocytic syndrome. The laboratory and radiologic tests available for the diagnosis are nonspecific. The common laboratory abnormalities are leukopenia, usually neutropenia; anemia; thrombocytopenia; elevated C-reactive protein and erythrocyte sedimentation rate; impaired liver function; and atypical lymphocytes on peripheral blood smear.

This disease is misdiagnosed as malignant lymphoma in up to one-third of cases and is associated with the development of systemic lupus erythematosus (SLE) and other autoimmune diseases.The differential diagnosis is challenging as many other conditions such as malignant lymphoma, metastatic disease, tuberculosis and infectious lymphadenopathies can present in a similar way. KFD is considered a self-limiting disease: spontaneous regression may occur between 1–6 months (4). In more severe cases, nonsteroidal anti-inflammatory drugs (NSAIDs) and/or steroids treatment has proven beneficial.

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

Lymph node excissional biopsy, bone marrow aspirate and biopsy, short term central venous catheter positioning, liver biopsy, upper airway endoscopy or urgent tracheostomy.

Ultrasonographically guided biopsy is a profitable strategy, because it allows select the lymph nodes with less necrotic tissue to reach a histological diagnosis.

Other incidental surgeries apart from disease or for diagnosis may also be required in such children.

Type of anaesthesia

There is no definite recommendation for either general or regional anaesthesia. A moderate sedation, associated with local or regional anaesthesia, may avoid difficult upper airway management due cervical lymphadenopathy. Spontaneous breathing or CPAP assisted ventilation are suggested.

In order to limit airway management, any neuromuscular blocker should be avoided, as much as deep sedation or general anaesthesia.

To date, a role of anaesthetic drugs as trigger factor for KFD has not been reported in literature.

Necessary additional diagnostic procedures (preoperative)

Cardiac function tests (electrocardiography, echocardiography) according to general conditions (prolonged fever, malnutrition, dehydration, and severity of phlogosis indices' alteration).

Blood examinations, enlarged metabolic or coagulation tests.

BNP blood level is useful to monitor cardiac failure, if suspected.

X-ray of the thorax, lung ultrasound, and a CT scan to rule out other pathologies, define the extent of the lesion and to locate the most accessible adenopathy for the biopsy.

Consultation of a specialist to document for juridical reasons already existent deficits, e.g. of neurological nature.

Particular preparation for airway management

Patients with significant cervical lymphadenopathy could benefit from steroids therapies before and after surgery.

Since cervical lymphadenopathy of KFD has often been important, it requires to be trained on 'cannot intubate/cannot ventilate' protocol and ready with supraglottic devices, video laryngoscopy, airway endoscopy device, and cricothyrotomy set.

Consultation of an ENT specialist for more detailed examination if required.

Planning of a safe clinical pathway for induction of anaesthesia if airway management cannot be avoided.

Particular preparation for transfusion or administration of blood products

There are no particular recommendations for blood products or transfusion administration.

Particular preparation for anticoagulation

There is no evidence to support the need of particular anticoagulation.

Particular precautions for positioning, transport or mobilisation

Not reported.

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

Not reported.

Anaesthesiologic procedure

Induction of anaesthesia either administering endovenous or inhaling volatile anaesthetics are allowed. In case of serious upper airway narrowing due to cervical lymphadenopathy and expected airway management, an ENT specialist support is required. Therefore, a non invasive airway management is supposed as much as possible.

Perform steroid replacement therapy in anesthesia induction where appropriate.

Particular or additional monitoring

No particular monitoring is required.

In case of KFD with neurological involvement, BIS monitor or intraoperative EEG monitoring may avoid or prevent neurologic state worsening.

Possible complications

Upper airway swelling and obstruction related to difficult airway management for severe cervical lymphadenopathy.

Rare complications:

- Cardiac complications such as tamponade.

- Interstitial lung disease and pleural effusion.

- Hepatitis is another rare complication.

- Hemophagocytic syndrome.

Postoperative care

Possible ICU admission for post-operative monitoring or weaning of mechanical ventilation in expected difficult airway management.

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 disease

Related to actual upper airway obstruction.

Ambulatory anaesthesia

As described above for type of anaesthesia.

Obstetrical anaesthesia

As described above for type of anaesthesia.

Literature and internet links

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These guidelines have been prepared by:

Author
Emanuele Rossetti, Linda Appierto, Roberto Bianchi, Sergio Picardo, Anaesthesiologists, Pediatric Intensive Care Unit, DEA-ARCO Department, Bambino Ges u Children's Hospital, Rome, Italy
emanuele.rossetti@opbg.net

Peer revision 1
Diego F Baenas, Clinical Medicine Department, Hospital Privado Universitario de Córdoba, Naciones Córdoba, Argentina
baenashospitalprivado@gmail.com

Peer revision 2
Chung Ching, Hospital Nacional Arzobispo Loayza, Lima, Perú
jorgechung14@gmail.com