



Kaposiform Haemangioendothelioma & Tufted Angioma

■ What are kaposiform haemangioendothelioma & tufted angioma?

Kaposiform haemangioendothelioma (KHE) and tufted angioma (TA) are rare benign tumours of blood vessels.

Although non-cancerous, they can grow fast and quite extensively in the affected area.

KHE and TA is often associated with Kasabach-Merritt syndrome (see below).



Kaposiform haemangioendothelioma

■ How does kaposiform haemangioendothelioma & tufted angioma present?

KHE presents as a firm, deep nodule or swelling that has a bruise-like or purplish discoloration. It can grow or expand rapidly, especially during infancy.

TA presents as reddish nodules, swellings or patches that can grow over months.

When lesions of KHE or TA become hard and firm, it may mean that there is blood trapping within the lesion, leading to Kasabach-Merritt syndrome (see below).

■ What is the Kasabach-Merritt syndrome?

Kasabach-Merritt syndrome or KMS occurs when blood cells and proteins responsible for blood clotting are trapped within a blood vessel tumour, leading to an increased risk of bleeding.

In infants, KMS is most often caused by a growing KHE or TA.

Infants with KMS present with swelling of the blood vessel tumour, associated with features of bleeding, including bruises, nose bleeds (epistaxis), blood in the urine (haematuria) or blood in the stools (haematochezia).

How are these conditions diagnosed?

KHE, TA and KMS are diagnosed by a combination of clinical history, physical examination and tests including blood tests, ultrasound, magnetic resonance imaging (MRI) and tissue biopsies.

KMS can be confirmed by blood tests including full blood count and clotting profile.

Ultrasound is a non-invasive, painless test to aid in diagnosis of KHE and TA. It can be performed either in the clinic or at the diagnostic imaging centre. It involves using a probe placed on the skin over the site of the suspected lesion. Depending on the size, this may take a few minutes to 30 minutes.

MRI may also be utilised to aid in the diagnosis and assess the extent of KHE and TA. There is no radiation involved. However, the infant or child needs to stay still for about 30 to 60 minutes, rarely longer. General anaesthesia (GA) may be required for infants and younger children who are unable to cooperate. GA is administered by our team of paediatric anaesthetists.

Tissue biopsies may be required for the diagnosis of KHE and TA. If required, this involves cutting the skin and removing a small piece of tissue to look under the microscope. This procedure may be done under local anaesthetic if the child is cooperative. Otherwise, it can also be performed under sedation in the ward or under GA in the operating theatre.

How are kaposiform haemangioendothelioma and tufted angioma treated?

Treatment of KHE, TA and KMS require multi-disciplinary management by a vascular anomalies team.

Treatment options include infusion of blood products to stop or prevent bleeding, and systemic medications to shrink it, including corticosteroids (e.g. prednisolone), sirolimus or certain chemotherapy drugs like Vincristine.

Rarely, embolisation or radiation therapy may be required for treatment.

If the tumour is small and accessible, surgery may be done.

Useful telephone number

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