Kaposiform haemangioendothelioma

What is Kaposiform haemangioendothelioma?

Kaposiform haemangioendothelioma (KHE) is a rare vascular (blood vessel) growth that may involve the skin and/or internal organs. It usually appears at birth or soon afterwards and in the early stages can be confused with other types of birthmark, such as an infantile haemangioma (also known as a haemangioma of infancy). Although it may be referred to as a tumour, it is not cancerous and does not spread to other parts of the body.

KHE can be associated with a condition called Kasabach-Merritt Phenomenon (KMP). KMP is characterised by trapping of platelets in the growth which leads to a reduction in the ability of the blood to clot. As the growth gets larger, more platelets are trapped so the symptoms tend to get worse. The medical term for a low level of platelets is thrombocytopenia. Platelets are responsible for promoting clotting. If a child has too few platelets circulating in the blood, they might bruise more easily than usual or develop tiny pinprick bruises under the skin (called petechiae), or rarely can bleed excessively.

What causes KHE?

We do not fully understand what causes KHE. It probably develops early in pregnancy when the blood vessels are forming. It is a very rare condition. At GOSH, we have treated 30 children in the past five years.

We know that it is not an inherited condition – that is, it is not passed on from parent to child. It is also unlikely to have been caused by anything that was done during pregnancy. Males and females appear to be affected in equal numbers.

What are the signs and symptoms of KHE?

KHE involving the skin usually appears as a blue-purple area that may be flat or raised. It may be apparent at birth or soon afterwards. The skin over the growth may be shiny and stretched, and tender to touch. This most commonly affects the skin on the trunk or limbs, although it can occur elsewhere. If the growth is near a joint, it can cause pain and limit movement.

KHE arising in the abdomen or chest may not be evident until it has grown to a size leading to swelling and pain, and
compression of adjacent structures. This can be life threatening if the airway, heart or lungs are affected.

**How is KHE diagnosed?**

As KHE is such a rare condition, diagnosis is usually made at a specialist centre with input from different specialists, such as dermatologists (skin specialists) and haematologists (blood specialists). Initially, imaging scans such as magnetic resonance imaging (MRI) or ultrasound will be used to work out the size of the growth and define its edges. If the growth is large, it may show up on routine prenatal ultrasound scans.

Once the size and shape of the growth is known, a biopsy (small sample of tissue) will be taken to examine under the microscope in the laboratories. Regular blood tests will also be needed to check to see whether KMP is present.

**How is KHE treated?**

The options for treatment depend on whether KMP is present or not. If there are no clotting problems and the growth itself is not causing any pain or compression of vital structures, treatment may not be needed, as the growth may shrink without treatment. Children will need regular monitoring to ensure that problems are identified at an early stage.

If a child has severe platelet and clotting problems, treatment is usually with a medicine called vincristine. This is a cytotoxic (poisonous to cells) medicine that is often used in cancer treatment. It is used in children with KHE and KMP to reduce the size of the growth and to reduce the clotting problems. It is usually given directly into a vein (intravenously) through a central venous access device, usually a central venous catheter.

Corticosteroid medicine may also be prescribed alongside vincristine to further reduce the size of the growth. This can also be given through the catheter, but is usually given by mouth. Another medicine called rapamycin (also called sirolimus) has been used in patients with KHE and KMP with promising results.

Embolisation by an interventional radiologist might be suggested to block the blood vessels in the growth, with the aim of stopping it growing and improving the platelet count. Very occasionally, depending on the size and location of the growth, surgery might be suggested. Removing as much of the growth as possible can reduce clotting problems, but this is not an appropriate treatment option for all children due to the risk of excessive bleeding during surgery.

KHE can also cover quite a large area so surgery is not appropriate in these cases.

Whichever treatment is used, your child's platelet count will be monitored regularly. Platelet transfusions are not recommended as the platelets are trapped in the lesion leading to increase in its size, and little or no improvement in the platelet count.
What happens next?

KHE usually resolves (shrinks) as a child grows older, although there may be small parts of the growth left behind. These can be surgically removed if troublesome.

KHE with KMP can be a very serious disease, and unfortunately if the bleeding problems are not treated successfully it can be life threatening. Treatment is intensive, but is successful in most cases. Once the growth has resolved, it cannot come back in later life.

Further information and support

At Great Ormond Street Hospital (GOSH), contact the Birthmark Unit.

The Birthmark Support Group offers support and advice to parents of children with all types of birthmark, including Kaposiform haemangioendothelioma. Telephone their helpline on 0845 045 4700 or visit their website at www.birthmarksupportgroup.org.uk.

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