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Kasabach–Merritt syndrome

What is Kasabach–Merritt syndrome?

Kasabach–Merritt syndrome is characterised by the combination of:

- A rapidly growing vascular tumour
- Thrombocytopaenia (low platelets)
- Microangiopathic haemolytic anaemia (destruction of red blood cells)
- Consumptive coagulopathy (impaired clotting).

The blood clotting disorder results from platelets and other clotting factors from the blood being used up (consumed) within the tumour.

What causes Kasabach–Merritt syndrome?

Kasabach–Merritt syndrome is caused by kaposiform haemangioendotheliomas, tufted angiomas and sometimes other vascular tumours. Previously Kasabach–Merritt syndrome was thought to be caused by large infantile [haemangiomas](#) (also known as cavernous haemangiomas or capillary haemangiomas) however this is not the case.

Kaposiform haemangioendotheliomas

Kaposiform haemangioendotheliomas are typically solitary tumours which appear in the soft tissues of the limbs, head and neck or retroperitoneum. They usually are seen in infants less than 2 years of age, although cases have been reported in adults. They do not spread (metastasise) but can cause serious problems because of local growth, cardiac failure or the associated Kasabach–Merritt phenomenon. There are few reports of kaposiform haemangioendotheliomas without Kasabach–Merritt syndrome. Kaposiform haemangioendotheliomas usually regress with time but do not completely disappear.

Tufted angiomas

Tufted angiomas usually present before 5 years of age, although they can occur throughout life. They present as brown, red or purple areas of skin and are firm to touch. They are often painful. Spontaneous regression is unusual. Most tufted angiomas do not cause Kasabach–Merritt syndrome and metastasis is rare.

Investigations

Kasabach–Merritt syndrome may be suspected in an infant with a vascular growth and evidence of easy bleeding or bruising. Blood tests may include a full blood count and film, clotting screen and fibrin degradation products. MRI, angiography, and biopsy may be performed but will depend on the clinical condition of the infant.

What is the treatment of Kasabach–Merritt syndrome?

A number of specialists may be involved in the care of a child with Kasabach–Merritt syndrome, including paediatricians, dermatologists, haematologists, surgeons and radiologists. Administration of blood products to correct the coagulopathy may be ineffective. A number of treatments have been used for Kasabach–Merritt syndrome including vincristine and [systemic steroids](#), as well as interferon alpha, [cyclophosphamide](#), aspirin and dipyridamole, and radiation therapy. Embolisation and surgical removal may also be considered.

Treatment of Kasabach–Merritt syndrome can be difficult. It is estimated that it has a mortality of around 20%.

Draft 8 May 2007

Related information

References:

On DermNet NZ:

- [Haemangiomas](#)
- [Vascular skin problems](#)

Other websites:

emedicine:

- [Kasabach–Merritt Syndrome](#)
- [Kasabach–Merritt Syndrome](#)

Books about skin diseases:

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DermNet does not provide an on–line consultation service.

If you have any concerns with your skin or its treatment, see a [dermatologist](#) for advice.

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