Conservative management of the Kasabach-Merritt syndrome (cavernous haemangioma and thrombocytopenia)

A report of 2 cases

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Summary

Two neonates with the Kasabach-Merritt syndrome (cavernous haemangioma and spontaneous bleeding) are described. The first infant bled into a rapidly growing cervical haemangioma. A consumptive coagulopathy was found, and he required blood transfusions; no response to steroids was observed. Further treatment was conservative, and at the age of 2 years he is well and the haemangioma is shrinking. The second infant had a strawberry naevus on the thigh which started bleeding when he was 10 days old, a buttock mass and a mass in the foot, thought to be lipomas. No clear benefit was derived from steroids. Bandaging of the thigh prevented further bleeding, and he is well at the age of 10 months. Conservative management seems best in this condition, and steroids appear to be of value in approximately 40% of cases.


The Kasabach-Merritt syndrome, or thrombocytopenia associated with cavernous or capillary haemangioma, was first described by Kasabach and Merritt in 1940. Since then about 60 cases have been reported. Surgery (including splenectomy), irradiation, and corticosteroids have all been used in the treatment of the thrombocytopenia, but lately conservative management has become the treatment of choice. Two infants are reported in whom the condition was managed conservatively after steroid therapy had failed. In both cases the thrombocytopenia resolved spontaneously and the haemangiomas diminished in size with time.

Case reports

Case 1

At birth a full-term Black infant, otherwise normal, had a capillary haemangioma measuring 1 x 1 cm in the left posterior triangle of the neck. Two weeks later he was admitted with a massive swelling at the site, filling the whole posterior triangle, extending on to the cheek and elevating the earlobe. Anaemia and thrombocytopenia were present; the haemoglobin concentration was 5 g/dl and the platelet count 1 000/ml. The prothrombin index was 50%. Other investigations were negative. Packed cells and vitamin K were given immediately, and three further blood transfusions were required during the next few weeks. Steroids were started at 40 mg/d but there was no response, the platelet count remaining at 6 000/ml or less. Coagulation studies showed diminished factor V, factor VIII and fibrinogen and a prolonged partial thromboplastin time. A bone marrow study was inconclusive, although megakaryocytes were seen. A radioactive technetium scan showed that the
haemangioma was fed by both an occipital artery and the internal carotid artery. Apart from developing *Haemophilus influenzae* meningitis at 4 months, from which he made a complete recovery, the infant did well; when he was last seen at the age of 2 years the platelet count and haemoglobin concentration were normal and the size of the haemangioma had decreased from a maximum of 22 x 19 cm to 4 x 3 cm (Fig. 1).

**Case 2**

An infant was admitted with gastro-enteritis at 5 days of age. He had a strawberry naevus measuring 2 x 3 cm on the right calf, an enlarged right buttock due to a mass in the gluteus muscle (thought to be a lipoma), and an abnormal left foot with enlarged first and second toes and small haemangiomas on the fourth and fifth toes (Figs 2 and 3). At 10 days the haemangioma on the leg started bleeding. Pressure bandaging was applied, but blood transfusions were necessary. The platelet count was then found to be low and steroids and more blood were given, but there was no clear-cut response. More blood was given at 45 days; thereafter the haemoglobin concentration and platelet count were normal, and no further bleeding occurred. At 10 months the haemangioma was much smaller but the buttock mass and the large left foot caused concern and the infant was referred for a surgical opinion. It was decided to leave both lesions until he reaches the age of 2 years, when plastic surgery for the foot and removal of the lipoma will be considered.

![Fig. 2. Case 2. The haemangioma on the thigh and the small haemangiomas on the foot.](image)

![Fig. 3. Case 2. The abnormal left foot.](image)

**Discussion**

Haemangiomas are common benign tumours of childhood. They usually present shortly after birth, may be single or multiple, and are found in any organ. Terms such as giant haemangioma, cavernous haemangioma, capillary haemangioma and strawberry naevus seem to be interchangeable, and according to Illingworth it is not always clear which is which. The natural history of the tumour is of rapid growth, with involution between 5 and 7 years and often earlier. Very rarely it may become malignant and occasionally there may be bleeding, sometimes into the tumour itself, after minor trauma or following the onset of a bleeding diathesis, as in the Kasabach-Merritt syndrome. Congestive cardiac failure, associated with multiple arteriovenous shunts in haemangiomas of liver, spleen, lung, brain, etc. has also been reported.

The bleeding diathesis can be due to thrombocytopenia alone, thought to follow trapping of platelets in the spleen or the haemangioma. It is not known why the platelets sequestrate, but this may be because of abnormal flow, abnormal vascular channels or increased liberation of thromboplastic substances in the haemangioma. Some authors have described jaundice in the newborn, petechiae, ecchymoses, fibrinogen deficiency, deficiency of factors V and VIII, a prolonged partial thromboplastin time and a prolonged prothrombin time, as well as a cycle of clotting and then clot lysis. A deficiency in the niurature of megakaryocytes and the release of platelets from the marrow has also been suggested.

Treatment ranges from conservative management to radical surgical excision. Irradiation of the haemangioma has frequently been tried, but this usually results in scarring and recurrence. Injection of nitrogen mustard, other sclerosing solutions or silicone particles and cryosurgery have all been tried when ligation of the feeder artery or excision has not been possible. Recently pressure bandaging, which is non-invasive and does not produce scarring, has been utilized. Prednisone or prednisolone up to 40 mg on alternate days has been successful in some cases. In one large series of over 200 patients with cutaneous, giant and mixed haemangiomas followed up for 2-6 years, over 80% of tumours had regressed spontaneously by the age of 2 years. Ulceration and infection of the lesion seemed to increase involution. Ten of 18 patients treated developed staphylococcal septicemia or scarring severe enough to require skin grafting. Local recurrence, pain and continued growth had followed treatment with dry ice, injections, surgery and irradiation. The authors concluded that conservative management produced the best results. Another hazard associated with surgery has been disseminated intravascular coagulation; full coagulation studies should be undertaken if an operation is planned. Disseminated intravascular coagulation may be responsive to heparin, and the use of fresh-frozen plasma and replacement of deficient factors should be considered in addition to steroids.

These 2 cases are reported to show that careful conservative management, with blood transfusions as required, is successful in the treatment of the Kasabach-Merritt syndrome. It has resulted in a normal blood picture and a regression in the size of the haemangiomas in both our patients. We recommend an initial conservative approach as described, with pressure bandaging if the haemangioma is in a suitable site. Steroid therapy could be tried if there is associated thrombocytopenia, but it must be remembered that this may be effective in only about 40% of cases. Surgery and irradiation should be reserved for those cases where the size or position of the haemangioma is an immediate threat to life since its natural course is to diminish with time.
REFERENCES


Amniotic bands
A case report
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Summary

The aetiology of amniotic bands causing intra-uterine annular constrictions of the fetus is discussed and a case report is presented. Cautious use of amniocentesis in the first trimester appears to be advisable.


The monstrosity is contrary to nature, not contrary to nature taken absolutely but contrary to the most usual course of nature. Nothing in fact can be produced contrary to that nature, which is both eternal and essential.

Aristotle, 4th century BC

Annular constrictions and intra-uterine amputations are believed to be the result either of an embryological development defect of the germ disc, an endogenous cause, or a mechanical constriction band from early rupture of the amnion, an exogenous cause.

A third theory that they may be due to an exogenous superimposed disease process of unknown aetiology, as suggested by Stocks and Stocks in supporting Streeter's original theory, is discussed and a report of a case in which the aetiology was possibly hereditary is presented.

The importance of a detailed history as well as histological examination of the placenta, the cord, the attachment sites of the bands and the sites of embryological abnormalities of the fetus should be stressed in gathering information on this condition.

Radiographs of any affected limbs should also be taken and congenital abnormalities should be looked for. Attention is drawn to the increased use of amniocentesis in early pregnancy; attending staff should be alerted to these anomalies and to the possibility that this procedure may precipitate amniotic damage.

Chemke et al. give an incidence for annular constrictions of 1 in 5000 to 1 in 10 000 pregnancies.

Case report

An infant was born prematurely on 31 January 1979. Before delivery the gestational age was estimated as 32 weeks by means of ultrasonography, and no fetal abnormalities were detected. The mother was a 23-year-old White woman, para 0, gravida 1; her last menstrual period had commenced on 10 June 1978 and the expected date of delivery was 17 March 1979. She was initially seen at 20 weeks' gestation and again at 24 weeks when ultrasonography was carried out because the patient was large for dates and had polyhydramnios; twins were suspected but only a single fetus was found. The mother, a teacher, gave a history of rheumatoid arthritis since childhood and had been exposed to acetylsalicylic acid and unknown homeopathic remedies in the first 12 weeks of her pregnancy. There was no family history of congenital abnormalities, rhesus disease, diabetes mellitus or hypertension in pregnancy on the maternal side. The father, a 25-year-old draughtsman, gave a history of club feet in his mother and cleft palate in a first cousin.

On admission the patient was in premature labour; it was attempted to stop labour with intravenous hexoprenaline (Ipradrol) 50 mg/l, regulated to maintain the pulse rate at under 120/min, but a fetus with gross congenital abnormalities was delivered. The baby's Apgar score at birth was 8 but dropped to 2 after 5 minutes, and she died 6 hours later.

The upper lip and forehead showed deep facial fissures (Fig. 1). The upper and lower limbs were well formed, but some digits of the hands and feet had band constriction. There was a constriction of the right hallux and second and third toes of the left foot (Fig. 2), a constriction of the base of the first finger, and a missing index finger on the same hand. There was a deep