that in the long term and on the basis of the radiographic evidence our patient's bone structure may be normal.

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Received 1 August 1983

Haemangioma with thrombocytopenia (Kasabach-Merritt syndrome)

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SUMMARY We describe two patients with haemangioma with thrombocytopenia (Kasabach-Merritt syndrome). Both were treated with corticosteroids without notable improvement. The first patient responded satisfactorily to radiotherapy, whereas the second showed a slow spontaneous resolution.

Kasabach and Merritt¹ first described the syndrome of thrombocytopenia with haemangioma. Approximately 100 cases have been reported subsequently. The haemangioma enlarges, usually in infancy, and at the same time there is a fall in the platelet count, together with hypofibrinogenaemia and a consumption coagulopathy. Radioisotope studies have shown an increased uptake within the haemangioma of platelets or fibrinogen, or of both.² Surgery is not always possible. Radiotherapy, corticosteroids, anticoagulants, and antifibrinolytics have all been used, but none is clearly superior. We report two girls with haemangioma and thrombocytopenia in whom the use of corticosteroids was of little or no benefit.

Case reports

Case 1. The haemangioma presented at the age of 5½ weeks, and progressively enlarged. On admission at 7 weeks there was an extensive firm swelling below

the chin extending into the neck on both sides but more on the right where it extended to the angle of the jaw (Figure). The haemangioma was also visible on the floor of the mouth below the tongue. The liver was palpable 3 cm below the costal margin. Investigations showed haemoglobin 8.5 g/dl, white cells $7.9 \times 10^9/l$, and platelets $20 \times 10^9/l$. The blood film showed microcytosis, anisocytosis, burr cells, and a normal differential. The prothrombin time was 12.5 seconds (control 11.5 seconds), activated partial thromboplastin time 45 seconds (control 43 seconds), and fibrin degradation products between 10 and 40 $\mu g/ml$ (normal less than 10 $\mu g/ml$).

Treatment with epsilon aminocaproic acid and vitamin K was given for three weeks, followed by one week of prednisolone 4 mg/kg/day, but the haemangioma continued to enlarge rapidly and the platelet count fell to $9 \times 10^9/l$. Surgical removal was impossible. Diminution in the size of the lesion occurred after a single dose of radiation (400 rads), but this was followed by enlargement of the lesion and haematological relapse. A second dose of 400 rads was followed by slow and complete regression of the lesion, so that the lesion had entirely disappeared by 14 months of age.

Case 2. A red swelling on the medial side of the left knee, presented at the age of 3 months, diagnosed initially as cellulitis and treated with penicillin.

Figure Case 1, the patient aged 10 weeks.

The lesion settled. Two weeks later it recurred. The left knee and thigh were very swollen with a hot, red indurated area medially, and within a day there were signs of haemorrhage around the lesion. Investigations showed haemoglobin 10.3 g/dl, white cells $12.2 \times 10^9/l$, platelets $41 \times 10^9/l$, and the blood film showed occasional crenated cells with scanty large platelets and a normal differential. The bleeding time was greater than 9 minutes. The prothrombin time was 11 seconds (control 12 seconds), activated partial thromboplastin time 39 seconds (control 42 seconds), thrombin time 15 seconds (control 10 seconds), and fibrin degradation products 40 $\mu g/ml$ (normal less than 10 $\mu g/ml$). A radiograph of the leg showed no bony abnormality. After transfusion of two units of platelet concentrates the lesion seemed smaller, but the platelet count soon fell further and bleeding into the lesion recurred. Treatment with prednisolone 2 mg/kg/day was started. After four weeks the platelets remained low at $9 \times 10^9/l$. The prednisolone dose was halved. At 6 months of age the platelet

count started to rise, became normal over the subsequent two months, and the prednisolone dose was tapered off. The haemangioma slowly diminished in size. When last seen at the age of $7\frac{1}{2}$ years, the left leg was 0.2 cm longer, and there was faint skin staining at the site of the original haemangioma.

Discussion

Large haemangiomas without thrombocytopenia regress well with corticosteroids.^{3,4} Unfortunately, when thrombocytopenia is a complication, only 50% of cases respond to corticosteroids. There is no feature that enables early recognition of the case which is likely to respond. The principal disadvantage of corticosteroid treatment is the danger of delaying more effective treatment. Radiotherapy was the treatment used in the original report 40 years ago by Kasabach and Merritt.¹ It has been used, sometimes in conjunction with steroid treatment, on several subsequent occasions,^{5,6} and is successful in about two thirds of cases. The disadvantages of radiotherapy include local failure of growth; destruction of cartilage, bone, and teeth; and possible late malignancy. However, for the patient with an inoperable tumour (as in our two) it seems to offer a better prospect of cure than any other form of non-operative treatment.

We recommend an initial trial of corticosteroids in all cases which fail to settle. An inoperable case which shows no response within two weeks should be treated with radiotherapy.

We thank the Department of Medical Illustration at Booth Hall Children's Hospital, Dr D Pearson for arranging radiotherapy for patient 1, and to Mrs G Sanders for typing the manuscript.

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Received 10 September 1983