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GIANT HEMANGIOMA AND THROMBOCYTOPENIA IN A NEWBORN INFANT TREATED BY IRRADIATION THERAPY

Case report

by

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The simultaneous occurrence of hemangiomas and thrombocytopenic purpura is unusual. The first report of a case of this now generally accepted clinical syndrome was published by KASABACH & MERRITT in 1940, and more than 40 cases have since then been quoted in the literature. HILL & LONGINO (1962) described two cases of their own and reviewed earlier case reports.

The size of the tumour and the severe bleeding tendency in these patients frequently pose considerable therapeutical problems. The present case concerns a newborn baby with a rapidly growing hemangioma and severe thrombocytopenia in which good clinical improvement was obtained by roentgen irradiation therapy.

Case report

A boy born by spontaneous delivery after a full-term uncomplicated pregnancy; weight at birth 4 450 g; general condition good and skin colour normal. A large tumour, which appeared to be a hemangioma, was evident in the left thigh; no petechiae or other signs of bleeding were

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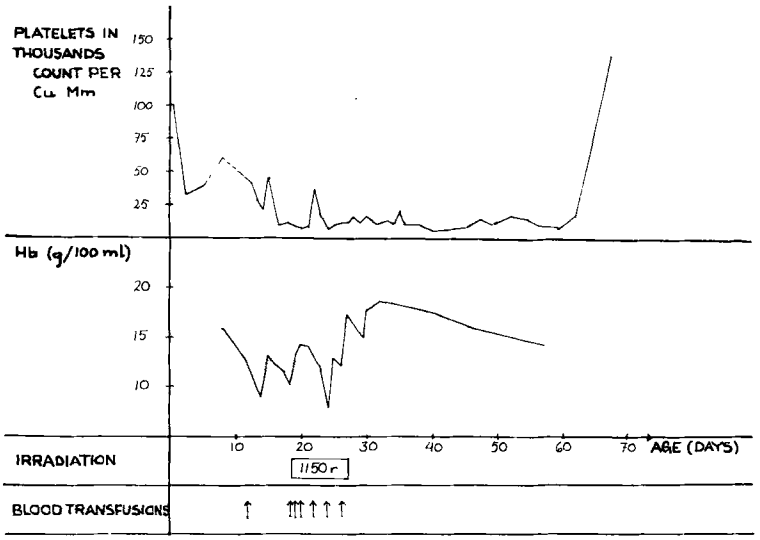

Fig. 1. Hemoglobin concentration and platelet count while in hospital. The days on which transfusion and roentgen therapy were given are indicated.

apparent. When the child was two days old the hemoglobin value was 16.7 g/100 ml and the thrombocyte count 99 000/mm³; on the fourth day the thrombocyte count had dropped to 33 000/mm³, and the child was transferred to the Childrens Clinic.

On admission the child's condition was still good and no signs of bleeding were evident. The liver extended 3 cm below the costal margin and the spleen was not palpable. An elevated hemangiomatous mass, purple in colour, firm and immovable, and 1.5 cm in size, was present over the right thigh, almost the whole of which was involved except for a small area posteriorly. The adjacent skin was not sharply demarcated from the mass. The leg appeared to be otherwise normal. The hemoglobin concentration was 15.5 g/100 ml, and the white cell count 14 300/mm³; the thrombocyte count was 59 000/mm³. The variations in hemoglobin concentration and platelet counts are depicted in Fig. 1. The prothrombin and bleeding times were normal. Tibial bone marrow aspiration disclosed a cellular marrow with slight myeloid hyperplasia and normal erythropoiesis; megakaryocytes were present and seemed to be increased in number though they were normal in form and possessed a granular cytoplasm. *Biopsy* suggested a capillary hemangioma with considerable endothelial proliferation.

Roentgen examination revealed only soft-tissue swelling of the thigh; the bone appeared to be intact. The tumour increased progressively in size during the following two weeks to form a circular mass that extended distally below the knee and proximally to the gluteal region (Fig. 2). There was marked edema of the leg.

From the age of two weeks the child bled continuously from numerous small ulcerations on the surface of the tumour; at the same time he gained as much as 100 g a day in weight, though this may be attributed in part to bleeding into the mass. The blood loss was considerable and sufficient blood volume was maintained by frequent transfusions of fresh blood; 800 ml in all were given (Fig. 1).

Irradiation therapy was started when the child was 18 days old. The tumour was cross-fired through two fields each measuring 8 × 10 cm; factors 180 kV, filter 0.5 mm Cu, FOD 40 cm, and HVL 1.1 mm Cu. A total of 1 150 r to each field was given on consecutive days in single

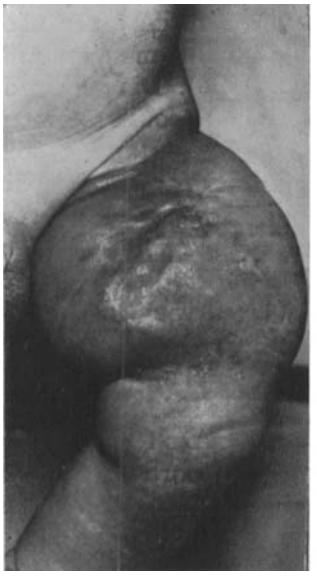

Fig. 2. Appearances of tumour prior to treatment.

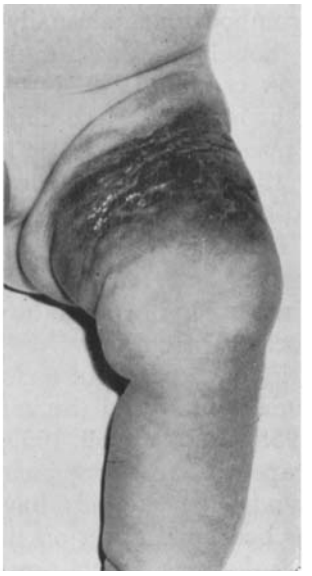

Fig. 3. The thigh at the control examination.

doses of 300 r with one of 250 r. A lead rubber sheet, 3 mm thick, was used to protect the gonads and epiphyseal lines; the secondary radiation caused by each single treatment (300 r) was measured as being 2.3 r to the gonads and 2.0 r to the nearest epiphyseal line.

The tumour felt much harder after a few days and there were signs of inflammation which included local elevation of temperature and maceration of the skin. The growth of the tumour was clearly inhibited and in spite of persistently low platelet counts the bleeding subsided towards the end of the treatment period. Clear improvement in the baby's general condition, which had been rather poor during the period of severe bleeding, was apparent at this stage. On the other hand there was no marked change in the size of the tumour, and it was not until 38 days after the treatment was completed that the platelet count rose dramatically and then remained normal (Fig. 1). The skin lesions also gradually healed, and the child was discharged after 67 days in hospital.

At control examination 3 months later, when the baby was 5 months old, his general condition seemed to be satisfactory. The tumour had almost disappeared, leaving a fibrotic scar formation, 5 × 10 cm in size and firm to the touch. There was no difference in the circumference of the thighs (Fig. 3). The peripheral blood picture was normal, as was the bleeding time. The thrombocyte count was 265 000/mm³. The growth of the femora appeared roentgenographically to have been equal.

Discussion

Thrombocytopenia and a tendency to bleeding in connection with a hemangioma is rare immediately after birth. Hemorrhage in this period constituted a problem in only 5 of reported cases (Hill & Longino 1962). The remaining cases reported were mostly in infants and children, with a few in the older age

groups. The thrombocytopenia usually appears during a period of rapid growth of the tumour, though cases with multiple small hemangiomas have also been described (GILON et coll. 1959, FISCHLER 1960). Diagnostic difficulties may arise when the hemangioma is situated subcutaneously or internally, e. g. in the intestine (JAMES & TUTTLE 1961, HILL & LONGINO 1962, BACKMAN & PARKKULAINEN 1958). Because of the tendency to severe bleeding, biopsy has been performed only in a limited number of cases and has revealed either a simple benign hemangioma, a capillary hemangioma or a hemangioendothelioma. Reports of bone marrow studies have not, however, recorded significant abnormalities except for immature megakaryocytes and a decreased or an increased number of these cells.

Several theories have been put forward to explain the process by which the thrombocytopenia is produced (SILVER et coll. 1948, SOUTHARD et coll. 1951, GOOD et coll. 1955, GILON et coll. 1959, BLIX & AAS 1961).

Various therapeutic measures, including supportive treatment, have been used in this syndrome. Steroids have occasionally been administered but without obvious beneficial effect on the thrombocytopenia (HILL & LONGINO 1962); they may, however, have prevented bleeding by raising the capillary resistance. Splenectomy has been performed in 8 cases with resultant improvement in the thrombocytopenia in one case (MEEKS et coll. 1955). Infusions of platelet concentrations restore the bleeding time to normal only for a very limited period but are of the greatest value before and during operative procedures.

The choice of treatment appears to lie between radical removal of the tumour and irradiation therapy. If the hemangioma is successfully excised there is a prompt rise in the platelet counts (INGLEFIELD et coll. 1961, HILL & LONGINO 1962). Operation is, however, ruled out in many cases because of technical difficulties or because of the great risk of excessive bleeding. Roentgen treatment results in cessation of the growth of the tumour and in its gradual shrinkage. The rise of the platelet count is slower than after surgery and occurs from one to several weeks after the treatment (SOUTHARD et coll. 1951, JAMES & TUTTLE 1961, SUTHERLAND & CLARK 1962). The period was about 5 weeks in the present case; on the other hand the bleeding from the tumour ceased much earlier when the treatment was completed, apparently as a result of the irradiation.

The irradiation dose was the usual one for hemangiomas and other benign tumours. The depth of the tumour was estimated to be approximately 3 cm, and a HVL of 1.1 mm Cu was used. The expected results were achieved: the growth of the tumour was checked, the thrombocyte count became normal and there were no signs of epiphyseal damage evident at the follow-up examination.

Disapproval of irradiation therapy is based upon the fear that secondary radiation may damage the gonads and epiphyseal lines. The total gonad dose in the present case was 18 r and 104 mr, and the dose to the femoral epiphyses

15 r and 320 mr, respectively. The therapy would appear to have caused no somatic damage but it is of course too early to evaluate possible genetic disturbances.

The writers believe that irradiation is indicated for cases with massive hemorrhages where radical treatment is impossible. A combination of irradiation and surgery might be advantageous; the acute stage could be controlled by radiation therapy and the tumour excised later (HILL & LONGINO 1962).

SUMMARY

A case of a newborn boy with a rapidly growing giant hemangioma and thrombocytopenia treated with roentgen irradiation is reported. The treatment resulted in a decrease in the size of the tumour and a rise in the platelet count. Some of the therapeutical aspects are briefly discussed.

ZUSAMMENFASSUNG

Ein Fall von einem neugeborenen Jungen mit einem rasch wachsenden Riesenhaemangiom und mit einer Thrombocytopenie behandelt mit Röntgenstrahlen wird beschrieben. Die Behandlung führte zu einer Verkleinerung des Tumors und einer Besserung in den Thrombocyten. Einige Punkte der Therapie werden erörtert.

RÉSUMÉ

Les auteurs présentent le cas d'un garçon nouveau-né atteint d'un hémangiome géant croissant rapidement et de thrombocytopénie, qui fut traité par roentgenthérapie. Le traitement amena une diminution du volume de la tumeur et une augmentation du nombre des plaquettes. Les auteurs discutent brièvement certains problèmes concernant le traitement.

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