GIANT HAEMANGIOMA WITH THROMBOCYTOPAENIA AND BLEEDING

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The earliest case of haemangioma with thrombocytopaenia and bleeding was described by Kasabach and Merrit (1940). The infant who was one week old had a haemangioma of the thigh, and eight weeks later, bleeding into the haemangioma was noticed with petechiae also on the skin. There was anaemia due to the bleeding, and the platelet count was 16,000 per cubic mm.

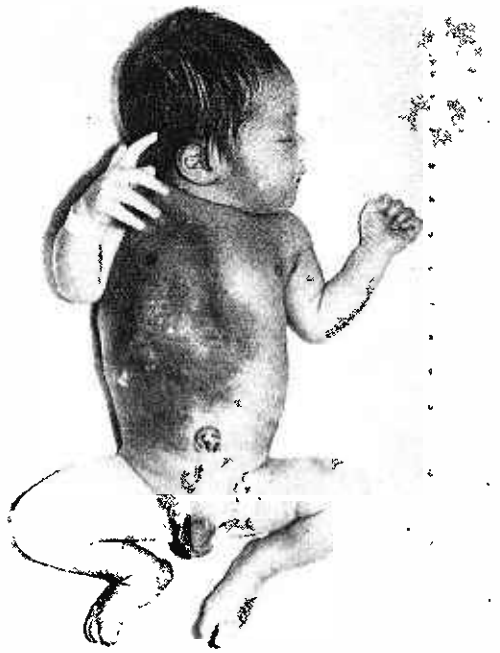
By 1959, there have altogether been 15 cases reported in English Medical Literature: (Rhodes et al (1944)), Silver et al (1948), Bogin et al (1951), Southard et al (1951), Franklin et al (1953), Weissman et al (1953), Good et al (1955), Meeks et al (1955), Dargeon et al (1959) and Gilon et al (1959). Therefore, although haemangioma with thrombocytopaenia is rare, this condition has been sufficiently reported to establish itself as a distinct entity. The following is a report of a case seen in the Paediatric Unit of the General Hospital, Singapore.

CASE REPORT

N.S.H. was born on 28th April, 1960 and he was seen by one of us (W.H.B.) to have a large cavernous haemangioma over the right hemithorax, extending from the midline in front to the midline at the back (Figs. 1a and b), and from the axilla to almost the level of the umbilicus below.

X-ray did not reveal any underlying bone involvement. A platelet count done at birth was normal, 200,000 per cub. ml. of blood, and except for the haemangioma, the infant was perfectly normal.

On the sixth day of life he was brought to hospital with a history of fever for one day. On examination, he had a temperature of 100°F. The haemangioma was warm and turgid and he also had petechiae all over the body. The platelet count done at this time was 50,000 per cub. ml. of blood, with normal bleeding, clotting and prothrombin times. The haemoglobin was 11.8G per 100 ml. of blood and the spleen was not palpable. The total white count was 8,900 with a differential count of 61% neutrophils, 30% lymphocytes, 3% eosinophils and 6% monocytes. Bone marrow aspiration showed the presence of



(a)

(b)

Fig. 1. Showing extent of haemangioma at age of 6 days.

normal megakaryocytes and a normoblastic hyperplasia. There were no abnormal cells. Because of the infected haemangioma, he was put on tetracycline and because of the thrombocytopaenia and bleeding, prednisolone 5 mgm. t.d.s. was also administered.

The next day he was bleeding from the umbilicus and from the sites of venepuncture, and the haemoglobin dropped to 5.9G per 100 ml. of blood, and a blood transfusion was given. This brought the haemoglobin up to 11G per 100 ml. of blood, but the patient was still bleeding. By this time the infection was controlled but thrombocytopaenia and bleeding into the haemangioma and at other sites were still present in spite of prednisolone therapy. One week later, the haemoglobin fell to 7.4G per 100 ml. of blood and another blood transfusion was given. In all, four blood transfusions were necessary to check the falling haemoglobin.

One month after admission, it was decided to institute deep X-ray therapy for the bleeding haemangioma. This was started on the 27th of June 1960. In view of the age of the patient and the thrombocytopaenia, treatment was started cautiously and controlled with daily blood counts. Two glancing fields were used to avoid irradiating the thoracic cage and its contents. Usually with deep X-ray therapy the haemoglobin and platelet count may fall but in this instance the platelet count rose and the haemoglobin which had been falling consistently, now was maintained at a high level. Altogether the total dose spread over a period of about three weeks came to 1000r.

Figure 2 shows very clearly the effect of deep X-ray therapy on the platelet count and haemoglobin level.

There does not seem to be any doubt that the radiotherapy had been responsible for the rise in platelets and cessation of bleeding. Since then the platelet count has been normal up to January 1961, and no more blood transfusions were required. At the same time, the haemangioma was shrinking in size due to the deep X-ray therapy. Figures 3 a and b show the

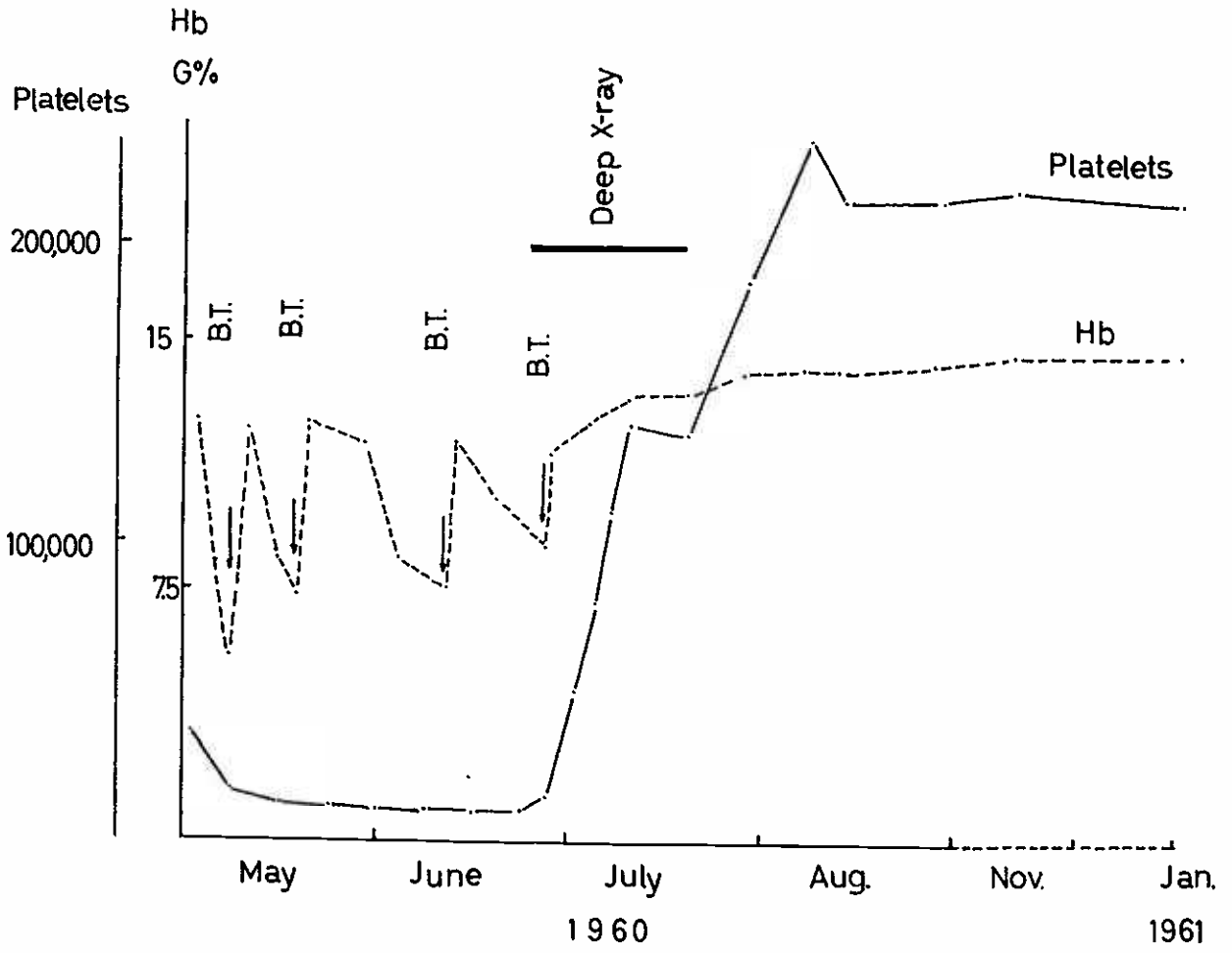
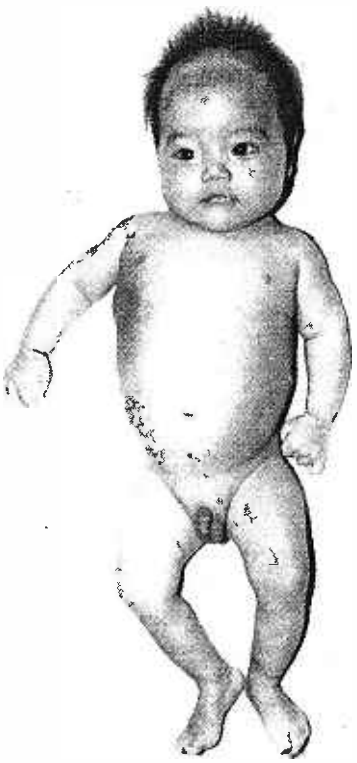
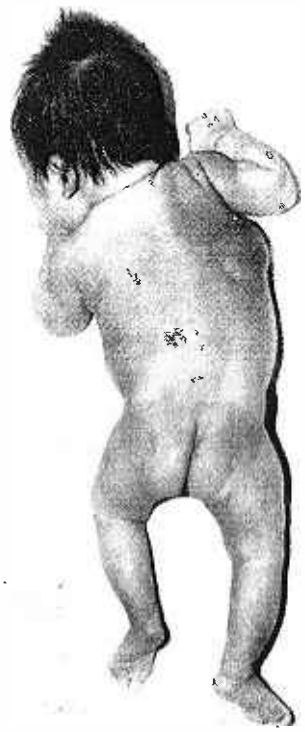


Fig. 2. Showing the low platelet count and falling haemoglobin in spite of blood transfusion, and the dramatic results of rising platelet and haemoglobin levels on deep X-ray of the haemangioma.

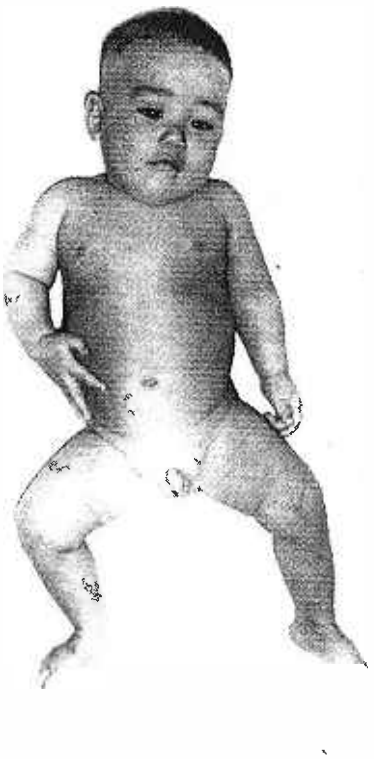


(a)



(b)

Fig. 3. Shrinkage of the haemangioma 2 weeks after cessation of deep X-ray therapy.



(a)

(b)

Fig. 4. Showing condition in January, 1961 with almost total disappearance of haemangioma.

extent of the swelling two weeks after the cessation of treatment.

The child has been followed up to now for a period of over six months. Figures 4 a and b show that at present the haemangioma has to all intents and purposes totally disappeared, and the child appears well and healthy.

DISCUSSION

It may be logical to assume that, since haemangiomata are common in infancy and that thrombocytopaenic purpura is also not uncommon the presence of both conditions in a patient may just be coincidental. But as the case above illustrates, the disappearance of the haemangioma resulted in the rise of the platelets and cessation of bleeding, and this satisfactory result is borne out also by many of the reports quoted above.

There have been various theories regarding the aetiology of this condition. Silver et al (1948) postulated that the following factors may be operative :—

(1) Increased phagocytosis of the platelets by the reticulo-endothelial tissues.

(2) Increased utilisation of platelets in an attempt to repair multiple capillary defects.

(3) Decreased production of platelets by the megakaryocytes in the bone marrow.

Good et al (1955) found in their case presence of platelet thrombi in sections of the haemangioma and considered that the thrombocytopaenia was due to the increased utilisation of platelets. Gilon et al (1959), compared the platelet counts in peripheral blood with the count from the haemangioma blood and found high platelet counts in the latter. In the present case, when the patient was first seen at birth, the platelet count was normal and when thrombocytopaenic purpura occurred on the sixth day of life there was evidence of bleeding in the haemangioma. The consensus of opinion at present is that the haemangioma is responsible for the mobilisation of platelets resulting in thrombocytopaenia and that the eradication of the haemangioma would result in a normal platelet count.

The management of this condition has been mainly centred on three procedures—splenectomy, administration of steroids, and irradiation of the haemangioma, together with supportive treatment. In Good's case (1955), splenectomy was carried out but the thrombocytopaenia persisted and the infant died five months after operation from fulminating infection. In one of Dargeon's cases (1959), splenectomy also was done and the platelet count was still low six months after operation. In another of his cases, steroids were given and splenectomy carried out, but the thrombocytopaenia persisted and the platelet count rose to normal only after radium therapy to the haemangioma. In all the other reports, satisfactory results were obtained only after irradiation. In the present case, the patient was on prednisolone for one month without any effect and splenectomy was not considered in view of the poor results after operation. There seems to be no doubt that the treatment of choice in this syndrome is irradiation of the haemangioma.

Finally, all the cases reported have been in infants with the exception of Gilon's case (1959) who was 13 years of age.

SUMMARY

1. The first case of giant haemangioma with thrombocytopaenia and bleeding seen in Singapore is reported, and the good response to irradiation of the haemangioma is described.

2. The arguments in favour of this being a clinical entity are given, and a short review of the theories of aetiology and its management are given.

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