THALASSAEMIA PRODUCING LEG ULCER
(A case report)

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Summary

Haematologic conditions producing leg ulcers are very uncommon except in hemoglobin-S homozygos state. Thalassaemia group of diseases causing leg ulcer is thus considered to be rare. A case of hemoglobin-E thalassaemia with leg ulcer is presented in this article.

Case Report

An 18 years old Hindu female suffered from jaundice and splenomegaly since she was 7 years old. She was treated in a sister institution for a while where she underwent splenectomy. According to the patient it was performed to relieve her jaundice, which disappeared for 1½ years. The patient visited the Dermatology OPD of our hospital with the complaints of weakness, jaundice and ulcer (of 1 year duration). Her sister 4 years younger to her, died 3 years before, with jaundice. On examination the patient looked her age. There was malar prominence and slight depression of the bridge of the nose. Skin was muddy yellowish in colour. Pallor and icterus were evident on conjunctiva. An ulcer (Fig. 1) was present on the medial malleolus. It was 1½" × 1" in size, more or less dry and showed a central area of mild inflammation. The ulcer was superficial, not tender and not adherent to deeper structures or bone. There was no bony tenderness. Skiagram of this region showed no bony involement. A splenectomy scar was present. Liver was enlarged 2 fingers breadth below the costal margin and was firm. No other abnormality was detected.

Laboratory findings were as follows :

Haemoglobin 5.2 gm% (100% = 16.0 Gm%), P. C. V. 24%, R. B. Cs. microcytic and hypochromic, W.B.C. 10.300/cmm (corrected for normoblasts), with neutrophil 55%, lymphocyte 43%, eosinophil 2% Platelets were present in sheets, Normoblasts 1500/100 leucocyte, reticulocytes 18%, direct Coombs' Test negative, Alkali resistant haemoglobin 21%, Bilirubin 5.3 mgm% (Indirect 5.0 mgm%), Haemoglobin electrophoresis in paper and cellulose acetate membrane showed feature of haemoglobin E thalassaemia.

Comments

Incidence of leg ulcer in cases of haemoglobinopathies is rare except in sickle cell anaemia where such incidence is known to be 5% - 75%[1]. In sickle cell thalassaemia the incidence of leg ulcer is 0 - 27%[2]. Leg ulcer is known to be rare in thalassaemia group of

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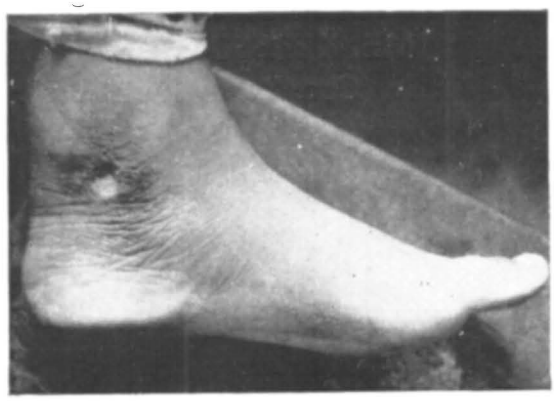
diseases. In thalassaemia major it has been stated to be very uncommon while in thalassaemia minor it does not occur at all. In about less than 5% of intermediate forms of thalassaemia leg ulcers may occur[3-4]. Till 1977, only 16 cases of leg ulcers in halassaemia have been reported in the literature[5]. The leg ulcers are possibly caused by inadequate tissue oxygenatino due to a lowered concentration of haemoglobin A. In our case foetal haemoglobin concentration was 21%. Lack of bony involvement due to this ulcer and lack of improvement by conventional therapy substantiates the haematologic etiology. Presence of excess of normoblasts and platelets in the peripheral blood film are consistent with post-splenectomy features. Mild indirect reacting bilirubinaemia and high reticulocyte count suggest the haemolytic features of this disease.



Picture of left ankle with foot showing the ulcer

Splenectomy in treatment of thalassaemia is known to bring about doubtful clinical improvement[6]. In a report of 3 cases of thalassaemia with leg ulcer, one showed clinical improvement following splenectomy. Another case who had splenectomy earlier developed leg ulcer later like in our case. In the third case there was improvement with conservative local therapy, even with the spleen extending down to the pelvic brim[6]. In our case where splenectomy was performed 11 years earlier, there was a temporary period of clinical improvement for $1\frac{1}{2}$ years. The operation did not however prevent the appearance of the ulcer.

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