MASSIVE THORACIC EXTRAMEDULLARY HAEMOPOIESIS IN A CASE OF HAEMOGLOBIN E THALASSAEMIA

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Extramedullary haemopoesis is not a common observation in the adult human body. In severe disturbances of haemopoietic function microscopic foci have been seen in the spleen, lymph nodes, and according to Wintrobe have been reported to occur less frequently in the liver, the suprarenal gland, in cartilage, in the broad ligament, in organising thrombi and in adipose tissue in various locations. Massive deposits simulating tumour growth are even less frequently encountered. These have been reported to occur in the paravertebral gutter, in the costovertebral angles and even subpleural nodules have been seen. Thus haemopoietic tumours have been found in association with erythroblastic anaemia, pernicious anaemia and other macrocytic anaemias especially that of liver disease, osteosclerosis and myelofibrosis, in cases of bone marrow replacement (e.g. carcinomatosis, Hodgkin's disease, erythraemia) hypoplastic anaemia, leukaemia and haemolytic anaemia. It is not clear why these massive extramedullary haemopoietic tumours occur in such curious sites, e.g. the paravertebral gutter. In cases where autopsies have been performed, no direct extensions from the marrow of adjacent ribs or vertebrae have been found. One view is that these tumours may arise from embroyonic haemopoietic tissue which has persisted into the post-foetal period and undergone hyperplasia under the influence of a prolonged stimulus.

A case of Haemoglobin E Thalassaemia with large paravertebral masses is reported.

CASE REPORT

E.C.N., a Chinese housewife was admitted into the Singapore General Hospital on 5th September 1961. She complained of one month's cough with breathlessness, vague chest and abdominal pain, stress incontinence of urine and marked weight loss. There was no significant past history but on direct questioning she admitted that she had been noted to have a yellow skin for at least the previous 20 years. There had been six pregnancies, three ended in miscarriages, two died soon after birth and only the fourth is alive and well. No details regarding the parents or siblings were available from the patient as they are estranged.

Physical examination revealed an ill-looking woman, who was dyspnoeic, mildly icteric and fairly severely anaemic. Signs of bilateral basal pleural effusion and a pericardial rub were noted and there was quite marked hepatosplenomegaly. No lymphadenopathy, ascites or oedema were present. Rectal examination revealed no abnormality. Vaginal examination revealed a retroverted uterus and chronic cervicitis.

Investigations: Hb. 6.96 gm%, W.B.C. 5,800/c.mm., Differential count: Neutrophils 74%, Lymphocytes 24%, Eosinophil 1%, Monocyte 1%, Reticulocytes 4.0%. The peripheral blood picture showed slightly hypochromic red cells, anisocytosis and polychromasia. Normoblasts and target cells were seen. Blood films for malarial parasites were negative. Blood for the L.E. phenomenon was negative.

Serum proteins: Total 7.9 gm%, Albumen 3.9 gm%, Globulin 4.0 gm%. Serum bilirubin: 1.0 mg%. Direct Coomb's test negative. Haemoglobin electrophoresis revealed the presence of Hb. "E" and Hb. "F". Blood urea 23 mg%. E.S.R. 24 mm.

Urine: Albumen present; M.E. No abnormality; Bile and urobilin trace; Urobilinogen+.

Chest aspiration: straw coloured pleural fluid withdrawn was negative on culture for pyogenic organisms and M. Tuberculosis. Direct smear showed R.B.Cs ++, lymphocytes ++, polymorphs + and few endothelial cells. Sputum for A.F.B. repeatedly negative on direct smear.

X-ray chest showed an oval opacity in the right base medially. A moderate opacity was seen in the left base, partly fluid. A further oval opacity was noted over the right superior

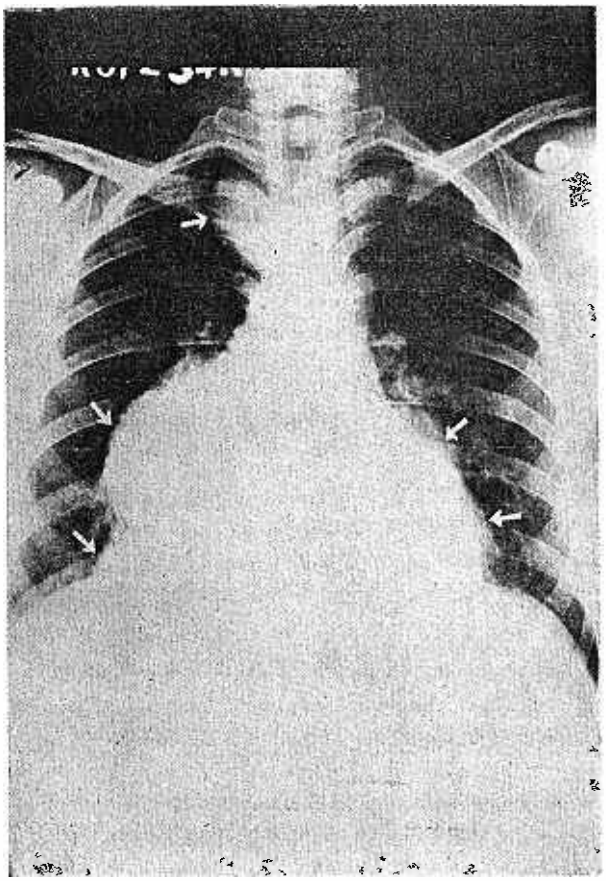

Fig. 1a.

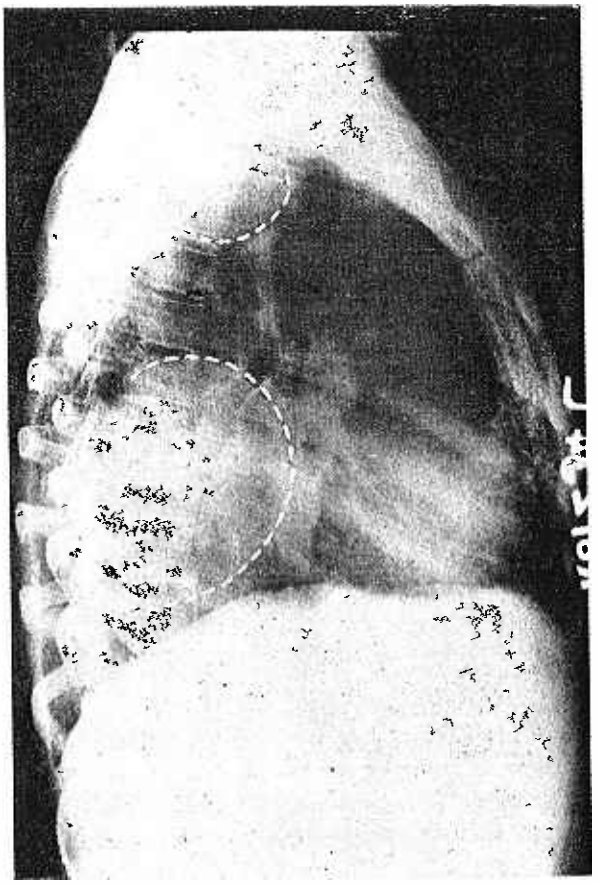

Fig. 1b.

Figs. 1a & 1b. P.A. and lateral X-rays of the chest showing the paravertebral masses.

mediastinum. The heart was moderatly enlarged. X-ray abdomen showed small opacities in the right upper lumbar region,, probable calcified gall stones.

Diagnosis and Progress: An initial diagnosis of Haemoglobin "E" Thalassaemia was made and probable reticulosis to account for the lung lesions. Without any specific therapy the patient's symptoms have cleared. However, the signs remain unchanged up to date, now 2 years since the patient first presented. A recent chest X-ray in August 1963, (Figs. 1a & 1b) shows a clearing of the pleural effusion. The heart remains enlarged and the rounded masses of varying sizes are still seen in both paravertebral gutters.

COMMENT

In view of the fact that the symptoms cleared without specific therapy and that the rounded shadows seen in the chest X-ray have remained unchanged over a period of 2 years it was felt that the chest lesions were benign and associating them with the haemolytic anaemia, they were probably extramedullary masses of haemopoiesis. Further proof of this was not possible as the patient was unwilling to undergo any operative procedure. The unusual feature in this case was the perticarditis and the pleural effusion.

SUMMARY

A case of Haemoglobin "E" Thalassaemia is described with pericarditis, a pleural effusion and radiological evidence of paravertebral masses in the chest which are being presented as probable extramedullary haemopoietic tumours.

ACKNOWLEDGEMENTS

I am indebted to Professor T. J. Danaraj for his advice and encouragement to publish this case.

REFERENCES

Dacie, J. V. (1960). The Haemolytic Anaemias. Part 1, 2nd Edition, Pg. 114.

Hanford, R. B., Schneider, G. F. and MacCarthy, J. D. (1960). Massive Thoracic Extramedullary Haemopoiesis. New England J. Med., 263, 120.

Hartfall, S. J. and Stewart, M. J. (1933). Massive Paravertebral Heterotopia of Bone Marrow in a case of Acholuric Jaundice. J. Path. & Bact., 377, 455.

Wintrobe, M. M. (1961). Extramedullary Haematopoiesis. Clinical Haematology, 5th Edition, 52.