SYSTEMIC MASTOCYTOSIS
(A case report)

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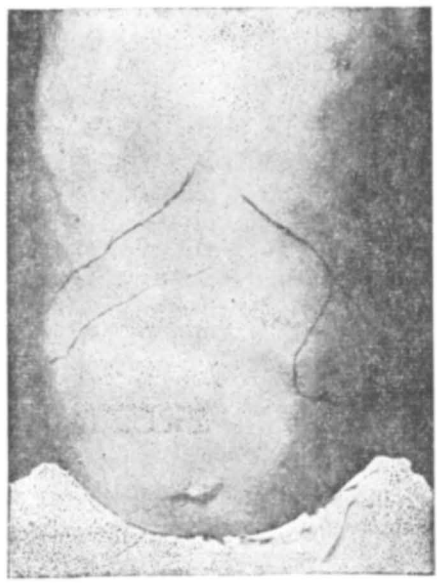
Summary

A case report of systemic mastocystosis with brief review of literature is presented.

Systemic mastocytosis is a rare disease characterised by abnormal proliferation of tissue mast cells. This condition was first described by Ellis in 1949[8]. Since then many case reports of systemic mastocystosis (Sagher[16], Berlin[2], Stark et al[17], Birt[3], Nickel[15]) have accumulated in the foreign literature. In Indian literature we came across only one report of systemic mastocytosis by Lahiri[13]. We are presenting report of a case of systemic mastocytosis in a male child.

Case Report

A 8 years old male child was admitted in the Skin and V. D. ward of Medical College and Hospital, Rohtak on 25—3—1970. His complaints at the time of admission were eruption of maculopapular lesions all over the body since the age of 2 months and distension of abdomen for the last 2 years. The lesions were first seen on the trunk at the age of 2 months and within about 3–4 months, they spread all over the body. The lesions turned red whenever they were scratched. He occasionally felt flushing of the face without any cause. The condition remained stationary for one year and then the lesions decreased in number. For the last two years abdomen of the patient had been gradually distending and the patient was aware of two masses in the abdomen (Figure). There was history of dyspepsia and occasional diarrhoea but no history of vomiting or generalised flushing was obtained. Patient was 4th in order of birth with 3 brothers and two sisters. Two brothers and one sister died due to some unknown cause. No history of similar condition could be traced in the family.



Papular lesions on the trunk. Liver and spleen have been marked out.

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On examination the patient was found to be moderately built and nourished. His B.P. was 110/80 mm. Hg. Axillary and cervical glands were palpable. Examination of chest C.N.S. and C.V.S. did not reveal anything abnormal. Liver was enlarged by 4 fingers. It was smooth, firm and nontender. Spleen was also palpable by 3 fingers and was firm and smooth.

Examination of the skin showed multiple, small (2mm–5mm) maculo papular lesions scattered mainly on trunk, face and genitalia but a few lesions were also seen on the limbs. The lesions were firm, waxy looking and shiny. A red flare was noted around the lesions immediately after they were stroked. Palms, soles and mucous membranes were free of any such lesions.

Investigations

Hb. 11 G.%, T.L.C. 4500cmm D.L.C. P42, L54, M–E4 E.S.R. 15mm. 1st hour. B.T. 3 minutes. C.T. 3.5 minutes. Platelet count. 90,000 cmm. urine N.A.D. Stools–N.A.D., Screening chest-N.A.D., S.T.S –negative, Serum alkaline phosphatase-10 K.A. units. Serum acid phosphatase-2 K.A, units, S.G.O.T.-38 I. U./L. S. G. P. T. 17 I. U./L., serum cholesterol 150mg., T.serum bilirubin 1.7mg%, Serum proteins–7.6g%, Thymol turbidity (½ hour) 2 units, Thymol Flocc. (18 hours)+++, X-rays of skull shoulder and hip joints-N.A.D. Skin biopsy: was consistent with the diagnosis of urticaria pigmentosa. Toluidine blue stain was used for mast cells. Collection of mast cells were seen under the epidermis. Liver biopsy: – Patient did not agree for liver biopsy.

Discussion

Though urticaria pigmentosa was described by Nettleship as early as 1869[14] the systemic nature of mast cell disease was recognised by Ellis in 1949[8]. He carried out autopsy on a child of urticaria pigmentosa and confirmed involvement of different systems by this process. Sagher[16], Clayman[14] and Stark et al[17] described bone lesions in cases of urticaria pigmentosa. Efrati[7], Friedman[11] and Szweda[18] described cases of fatal mast cell leukemias. Ende 1958[9] described a case of mastocytosis with involvement of spleen only. Lahiri[13] described 7 cases with involvement of liver, spleen, lymph nodes and bones.

Clinically the disease presents in different forms Demis[6] classified mastocytosis as under:

A: Benign (1) Cutaneous: Solitary-Mastocytoma Generalised Urticaria Pigmentosa.

(2) Systemic: Skin, bones liver, spleen, gut and lymph nodes.

(3) Splenic.

B: Malignant: Mast cell leukemia.

In our case we based the diagnosis of systemic mastocytosis on the findings of characteristic lesions of skin, positive Darrier's sign, involvement of liver, spleen and lymph nodes and history of dyspepsia, diarrhoea and flushing of face. Biopsy of skin proved the diagnosis.

Exact nature of the disease is still not known. Fischer 1962[10] was of the view that mastocytosis is an irreversible reticular proliferation. Hergeberzer 1959[12] thought that mastocytosis is a hyperplasia of highly differentiated elements of R.E. system. Lahiri 1966[13] holds that mastocytosis is a reticulosis which may be a link between cutaneo visceral malignancy.

Several modes of treatment have been advocated for mastocytosis. Urbach 1964[19] stressed the role of desoxycortisone in the treatment of mastocytosis. Davis 1958[5] advocated antihistominics as a therapy. Baer

1959[1] tried reserpine in cases of urticaria pigmentosa. In the present case we tried antihistaminics but no appreciable benefit was achieved.

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