CASE REPROT

URTICARIA PIGMENTOSA-A CASE REPORT

by

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Urticaria pigmentosa is a relatively rare disorder affecting the tissue mast cells of the body especially of the skin. In recent years, the concept of the disease has changed from a purely dermatological entity to a systemic disease. Even though review of foreign medical literature shows number of reports of urticaria pigmentosa (Dewar and Milne 1955[6], Nickol W. R., 1957[14] 1967[15], Davies etal. 1958[5], Brit and Nickerson 1959[2], there are very few reports in the Indian literature. This is a report of a case of urticaria pigmentosa in a female child.

Case History.- A two year old female child was admitted to the Dermatology Department of Medical College, Kottayam with complaints of multiple pigmented lesions on the body. The child was the second in the family and was born full term. At birth the baby had a mole like black area in the left infrascapular region. Later, multiple reddish brown areas appeared over the back, scalp and face. The mother gave a history of having had an acute attack of urticaria following aspirin therapy.

On examination the child weighed 8 kg. The mental age and physical development were normal for her age. She was active, intelligent and co operative. Sytemic examination did not reveal any abnormality. Skin showed multiple, papular, pigmented lesions distributed all over the trunk (Fig. 1 & 2). The lesions became itchy and more prominent on rubbing. The lesion in the infrascapular area was raised and tender.

Investigations:— Total leucocyte count-8200/cmm. Differential count- N_{38} L_{52} E_8 M_2 and B_0 Haemoglobin-80%.

A punch biopsy of the skin lesion was done. Histopathological examination revealed accumulation of round and stellate shaped cells in the upper dermis. Staining with toluidine blue revealed metachromatic granules in the cytoplasm of these cells. Diagnosis.- Urticaria Pigmentosa (Fig,3).

Discussion Urticaria pigmentosa was first described in 1869 by Nettleship[13], even though the name was given by Sangster in 1878[17]. The relationship between mast cell and urticaria pigmentosa was demonstrated by Unna in 1887[19] and during the period 1925 a series of reports and statistical

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compilations on urticaria pigmentosa appeared in the literature. Before World War II cases of urticaria pigmentosa used to be classified under two groups (Wieber and Hellerschmied 1930[20] ie. 1. congenital macular type in infancy and childhood and 2. acquired telengiectatic type in adults. Other types described are the xanthelasmoid type (Fox 1875 (9)) and the bullous type (Fox 1883 (10), Little 1905[12]a, 1906[12]b, Finnerud 1923[8], and Dewar and Milne 1955[6]). Systmic manifestations like flushing (Birt and Nickcrson 1959 [2]), lymphnode enlargement and hepatosplenomegaly (Ellis 1949 [7]), Berlin 1955[1]), (Nickel 1967[15]) and bone lesions (Sagher etal. 1952[16]), Starketal 1956[18] have been described. These are encountered mostly in patients in whom the disease starts in adult life and only rarely in infants. It is thought that the bone lesion is due to the collagenase effect of heparin of mast cells, and osteoporosis has occured in individuals to whom heparin has been administered (Griffith etal. 1965[11].

Nickel (1957 (14)) considered the various manifestations of urticaria pigmentosa and suggested that the finding of a cellular band in the upper third of the dermis indicates the need for special stains for mast cell.

Caplan 1963[4] classified the lesions into 3 groups:—

Group I Solitary.

Group II multiple, begining in infancy, and

Group III multiple, begining in early adult hood. He stated that Group II is the most common. Brett etal (1967)[3] reported 11 cases of mast cell disease in children and in 6 of these the lesions were present at birth. The lesions were maculopapular in 5, maculonodular in 2, and as isolated nodules in 4.

The present case belongs to Group II of Caplan (1963)[4]. The lesions were present at birth and they were maculopapular and this is in conformity with other cases of urticaria pigmentosa reported in infants (Caplan' 63,4) Bratt et al 1967[3] The mother of the child gave a history of urticarial rashes after taking aspirin tablets. Allergy is thought to be an important etiological factor in Urticaria Pigmentosa and Caplan (1963) (4) reported that a significant number of patients or their close relatives give a history of allergic disorder. Brett et al (1967) (3) reported an allergic family hisory in 4 out of 11 cases studied by him.

Cases of urticaria pigmentosa where the lesions appear in childhood and are confined to the skin undergo spontaneous resolution during adolescence unlike the adult form which is likely to persist indefinitely.

Summary:- A case of urticaria pigmetosa in a two year old female child is reported The literature is briefly reviewed.

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REFERENCES

1. Berlin C. Urticaria Pigmentosa as a systemic disease. Arch. Derm. 71,703, 1955.
2. Brit A. and Nickerson M. Generalised flushing of the skin with urticaria pigmentosa, Arch, Derm, 80,311 1959.
3. Brett E. D. M., Beale H. and Friedman T. Mast cell diseases in children; Brit. Jr. of Derm. 79, 197, 1967.
4. Caplan R. M. Natural course of urticaria pigmentosa, Arch. Derm. 87, 146, 1963.
5. Davies M. J., Lawler J. C. and Higdon R. S. Studies on an adult with urticaria pigmentosa-report or case. Arch. Derm 77, 224, 1958.
6. Dewar W.A. and Milne J. A. Bullous urticaria pigmentosa-summary of literature and report of two cases. Arch. Derm. 71, 717, 1955.
7. Ellis J. M. Urticaria Pigmentosa-report of a case with autopsy. Arch. Path. 48, 426, 1949.
8. Finnerud C. W. Urticaria Pigmentosa with summary of literature. Arch Derm. & Syph. 8, 344, 1923.
9. Fox. T. 1875 Quoted by Nickel 1967.
10. Fox. T. C. 1883. Quoted by Nickel 1967.
11. Griffith G. C. Heparin Osteoporosis J. A. M. A., 193, 91, 1965.
12. (A) Little 1905 A Contribution to study of Urticaria Pigmentosa, Brit. J. of Derm. 17, 355, 1905.
    (B) Little. A contribution to study of Urticaria Pigmentosa, Brit. J. of Derm. 18, 16, 1909.
13. Nettleship E. Rare forms of Urticaria-B. M. J. 2, 323, 1869.
14. Nickel W. R. Urticaria Pigmentosa-Mastosis. A consideration of various manifestations Arch. Derm. 76, 476, 1957.
15. Nickel W. R. Urticaria Pigmentosa-Mastosis-Arch. Derm. 96; 364, 1967.
16. Sagher F., Cohen C. and Schorr S Concomittant bone changes in urticaria pigmentosa-J. of Invest. Derm. 18 425, 1952.
17. Sangster. An anomalous mottled rash accompanied by pruritus, factitious urticaria and pigmentation. Urticaria Pigmentosa? Trans. Clin. Soc. Lond. 11, 161, 1878.
18. Stark E., Van Buskirk F. W. and Daly J. F-Radiologic and Pathologic bone changes associated with urticaria pigmentosa-Arch. Path. 62, 143, 1956.
19. Unna 1887-Quoted by Brett etal. 1967.
20. Weber F. P. and Hellenschmied R. Telengiectasia Macularis Eruptiva Perstans. Brit. J. of Derm. 42, 374, 1930.