MYCOSIS FUNGOIDES - TUMOUR D'EMBLE'E
(A case report with review)

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Summary

A case of mycosis fungoides-tumour d'emble'e-is reported and discussed. A leukemoid reaction or transformation of mycosis fungoides to other forms of reticulosis is suggested as the probable cause for raised W.B.C. count in our patient.

Alibert in 1806[1] first reported mycosis fungoides. He coined the term "Mycosis Fungoides" for this tumour because of its mushroom like appearance and not to indicate any fungal cause. This is a relatively rare, clinically easily definable, chronic itchy dermatosis which runs a progressive course with infrequent remission affecting primarily and predominantly the skin and in some cases the internal organs[2,3,4]. Although the nosological position of mycosis fungoides has not yet been firmly established, the name is still retained because of its historical precedent. Bazin[5] described this disease as being characterised by sequential development over a period of months or years of scaly patches, infiltrated plaques and ultimately tumours — the premycotic, infiltrated plaque and the tumour stages. The condition is generally considered to be a lymphoma.

Vidal and Brocq[6] were the first to describe the most disputed form of mycosis fungoides, the "tumour d'emble'e" form, which is characterised by development of tumour stage without any preceding patches or plaques. The tumours develop suddenly on an apparently normal looking skin. According to Lapiere[7] the development of a emble'e form is the result of limitation of pathological process to small areas of dermis. Whereas in premycotic stage lesion involves only the papillary dermis, in infiltrated stage it forms homogenous regions giving rise to thick plaques. A sudden expansion to the entire superficial dermis will give rise to the development of erythrodermic variety.

To our knowledge only less than 50 case reports of this rare form of mycosis fungoides are published in the world literature. The largest series of 40 cases was collected and published by Blue farb[2]. Among the remaining, 5 cases have been reported by Black et al[4], one case each by Saint-Andre et al[8], and Singh and Shah[9] and 2 cases by Singh et al[10]. Only 4 cases have been so far reported from India, one case each by Basu et al[11] and Singh and Shah[9], and two cases by Singh et al[10].

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All 4 cases were from North India. The authors are not aware of any cases of mycosis fungoides, d'emble'e form reported from South India especially from Kerala and hence this case report.

Case Report

On 30th September, 1974, a 40 year old housewife admitted to the Skin and V. D. Department of Medical College Hospital, Alleppey with multiple nodular swellings all over the body of one year's duration. The disease started with mild fever followed by the appearance of 2 or 3 swellings in the axilla, abdomen and face. Prior to admission she also had ayurvedic treatment and treatment from various taluk headquarters hospitals for the same complaint, but got no relief. She had also got admitted to one of the leprosy sanatoria with a suspicion of having lepromatous leprosy, which was ruled out later. Except for the swellings she had no other complaint during admission to the sanatorium. Meanwhile the nodular swellings increased in size and and number within a period of about 3 to 4 months they spread all over the body. Occasionally during the above period she used to get mild fever and cough with expectoration which used to subside with symptomatic treatment. 8 months after the onset of the swellings, she attained menopause, after which lesions increased suddenly in number and size. Patient never experienced any pruritus during the course of the disease.

Past history, family history and personal history were non-contributory.

General examination revealed an apparently healthy middle aged woman without any noticeable abnormality other than the swellings. Examination of skin showed multiple erythematous nodular swellings all over the body predominantly distributed over the face, neck, extensor aspect of forearms, lateral aspects of the elbows and dorsa of hands. (Fig. 1 Page No. 47) Individual lesions were either round or irregular in shape with a size varying from 1.5 to 2.5 cm. They were pink in colour, not scaly and majority of them were well circumscribed, discrete and without any surrounding lesions. They were neither infected nor tender. Few of the the lesions situated on the extensor aspect of hand and near the elbow showed smaller lesions around them with hyperpigmentation and scaling. Intervening skin appeared normal.

There was no significant lymphadenopathy. Systemic examination did not reveal hepatosplenomegaly or any other abnormality.

Investigations

Total Leucocyte count was 28,200 per cmm. with polymorphs 28%, lymphocytes 70% and eosinophils 2%. R. B. C. count was 4.8 million per cmm. Haemoglobin 8.2 gms%, E. S. R. 60 mm. after first hour and platelet count was 1,46,000 per cmm. Serum proteins, total 7.2 gms% with Albumin 3.2 gms% and globulin 4 gms%. Stool examination revealed presence of round worms. Urine examination did not reveal any abnormality. Bleeding and clotting time were within normal limits. S. T. S. (V. D. R. L.) was non reactive. Plain x-ray chest and abdomen and E. C. G. revealed no abnormalities.

Histopathology

One of the nodules from the extensor aspect of the hand was excised for histopathological examination. This revealed patchy areas of atrophy, acanthosis and elongation of rete pegs. Dermis showed a pleomorphic infiltrate composed of histiocytes and large number of cells with hyperchromatic nuclei. Some of the cells showed a prominent vesicular nucleus. Numerous mycosis cells with indented or

angular nucleus were also seen. The infiltrate was seen predominantly around blood vessels and at certain areas it extended to the subcutis. The picture was consistent with mycosis fungoides.

Treatment

After admission, she was given parenteral iron for 7 days and other supportive therapy and haemoglobin level was brought up to 13.5 gms%. Simultaneously she was given broad spectrum antibiotics to control the secondary infection which developed after admission to the hospital. Betamethasone cream (Betnovate) was given as external application.

After confirmation of diagnosis, patient was given a total 4.5 gram cyclophosphamide (Endoxan) by intravenous route, 100 mg. on the first day which was increased to 200 mg/day. Simultaneously she was also given 40 mg. of prednisolone per day. During the endoxan therapy she developed respiratory infection which was treated symptomatically. On the 6th day of endoxan therapy patient became restless and complained of pain and burning sensation all over the body especially over the lesions. This was controlled by external application of betamethasone cream, tranquilisers and sedatives.

Patient was discharged on 4—12—74. At the time of discharge the nodules on the extremities and trunk had subsided completely leaving hyperpigmented macules. The nodules on the face also had subsided remarkably, leaving some amount of residual infiltration at the site of nodules. Patient had no other clinical symptoms. The blood picture at the time of discharge showed a total leucocyte count of 5100 per cmm., with polymorphs 66%, lymphocytes 30%, eosinophils 4%, haemoglobin 12.5 gms% and E. S. R. 16 mm. after 1 hour.

Patient came for regular check up for subsequent 7 months. She was last seen in July, 1975. At that time she had no complaints except for mild infiltration of the face and a general examination revealed no abnormalities. She did not turn up for further follow up but was reported to have died suddenly in August, 1975.

Comment

Although Symmers[12] considered mycosis fungoides as a clinically and pathologically nonexisting entity, it is now accepted by majority of workers, as a well recognised dermatological entity. Cawley et al[13] and Clendenning et al[14] are of opinion that mycosis fungoides is only a clinical entity and not a histopathological one. Various schools hold different concepts about it. Opinion as to whether tumour d'emble'e form of mycosis fungoides represents a nosologic entity, is still more fluid.

Lever[16] describes the classical histopathological changes of mycosis fungoides as, "a great multiplicity of cell types, pleomorphism of histiocytes, presence of mitotic figures, presence of immature and atypical reticulum cells called mycosis cells, presence of patchy infiltrate in lower dermis and the presence of Pautriers microabscess in the epidermis".

In the present case we observed multiplicity of cell types, pleomorphism of histiocytes, presence of mitotic figures and patchy infiltrate in lower dermis, extending in some places to subcutis. These histological findings are consistent with those described by Lever[15]. Patchy areas of atrophy of the epidermis observed in this case can be attributed to the pressure exerted by the large amount of infiltrate on the epidermis. Limitation of pathological process in some areas of dermis especially around the blood vessels and appendages of the dermis can be explained on the basis of postulations by Lapiere[7]. According to him the development of tumour d'emble'e form

of mycosis fungoides is due to the limitation of pathological process to small areas of dermis.

Ormea and Pino[16] observed that in the tumour stage the overall appearance is one of intense cellular activity with an uneven nuclear staining, mainly hyperchromatic with variability in nuclear size and shape, which were observed by us in the present case also.

An interesting observation seen in the present case is the absence of pruritus. Presence of moderate to severe amount of pruritis is considered an essential manifestation of mycosis fungoides by majority of the workers. However Samman[19] has described the presence of asymptomatic lesions in mycosis fungoides.

Another interesting observation seen in the present case is the high leucocyte count with a predominance of lymphocytes. Vanscott and Haynes[17] described a normal blood picture in cases of mycosis fungoides. According to them the R. B. C. morphology and W. B. C. count are usually normal with a moderate eosinophilia of 6 to 12% in some patients. Leutner and Jordon[18] reported a marked elevation of W. B. C. count above 20,000 C.M.M. with majority of atypical monocytoid cells in the Sezary variant of mycosis fungoids which is characterised by intense pruritis, erythroderma with large monocytoid cells in blood and skin. Samman[19] also has described the development of mycosis fungoides from Sezary reticulosis. Krishnadas et al[20] has described the development of leukemoid reaction in malignancies due to nonspecific stimuli. Stratton[21] reported development of leukemoid tumours in mice after the inoculation of saline emulsion of a portion of mycosis fungoides tumour. This finding has not been further confirmed. However the authors are of opinion that the increased leucocyte count with predominance of lymphocytes observed in the present case is probably due to leukemoid reaction occurring in mycosis fungoides.

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REFERENCES

1. Alibert JLM: Description des maladies delapean observes a '1' Hospital Saint Luis, Paris, Borris 1806, p. 157. (Quoted by 2)
2. Bluefarb SM: Cutaneous manifestation of malignant Lymphomas, Charles C Thomas Publishers, Springfield, Illinois, 1959 p. 109.
3. Allen AC: The Skin, 2nd Edition, Grune and Stratton, New York 1967. (Quoted by 19)
4. Black JB: Edge Comb J, Eisen A et al: Mycosis fungoides natural history and aspects of its relationship to other malignant lymphomas, Amer J Med, 34: 228, 1963.
5. Bazin PAE: Maladies de la pean observes a '1' Hospital Saint Luis, Paris, 1876 (Quoted by 17)
6. Vidal E and Brocq L: E tude surle mycosis fungoide, france Med 2: 946, 1885. (Quoted by 14)
7. Lapiere S: J Invest Derm 42: 101, 1964 (Quoted by 19)
8. Saint-Andre P et al: Mycosis fungoides with a tumour of sudden onset. Bull soc Franc Derm Syph. 72: 6, 1965.
9. Singh S and Shah: Mycosis fungoides, J Assoc Phys India, 13: 209, 1965.
10. Singh R, Pandhi RK and Arora JK: Mycosis fungoides - Tumour D'Emble'e. Indian J Derm Vener, 39: 216, 1973.
11. Basu PN, Bhaskara MT and Pandalai KG: Mycosis fungoides, Brit J Derm, 41: 50, 1929.

12. Symmers D: Mycosis fungoides as a clinical and pathological non-existant. Arch Derm and Syph, 25: 1, 1932.

13. Cawley EP, Curtis AC and Leach JEK: Is mycosis fungoides a reticuloendothelial neoplastic entity? Arch Derm, 64: 225, 1951.

14. Clendening WE, Brecher G, Vanscott EJ et al: Mycosis fungoides. Arch Derm, 89: 785, 1964.

15. Lever WR: Histopathology of skin, 4th edition, Pitman Medical Publication, Philadelphia, 1967, p. 752.

16. Ormea F and Pino F: Minerva Derm, 40: 1, 1965 (Quoted by 19).

17. Vanscott EJ and Haynes HA: Cutaneous Lymphomas, Dermatology in General Medicine, Ist Edn. Edit by Fitzpatric TB, Arndt KA, Clark WH et al, Blakiston Publication, New York. 1971. p 556.

18. Leutner MA and Jordan HW: Ultra structure of an abnormal cell in Sezary syndrome. Blood, 31: 719, 1968.

19. Samman PD: Reticulosis, In text book of Dermatology, 2nd Edn. Ed. by Rook A, Wilkinson OS, and Ebling FJG, Blackwell scientific publication, Oxford, 1972, p 1384.

20. Krishnadas KV and Thomas M: Frequency and clinical presentation of leukaemias in Kerala State, Ker Med J, 16: 406, 1976.

21. Stratton ER: Arch Derm Syph 48: 179, 1943 (Quoted by 19).

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Of the 3 types of cytoplasmic antigens which can be distinguished by their location in different layers of the epidermis, one is found in all keratinocytes, another only in the upper layers of the epidermis and the third only in the basal cells. Antibodies to epidermal cytoplasmic antigens are found in approximately 20% of normal persons and in 30–40% of persons with various cancers. Antibodies to the cytoplasmic antigens present throughout the epidermis has an increased incidence in persons with various malignancies. Antibodies to basal cell antigens are rare and are reported in patients with drug reactions, burns, pemphigus and pemphigoid.

Reference: Bystryn J: Epidermal antigens, Int J Dermatol, 16: 645, 1977.