LETTER TO THE EDITOR

Successful PUVA-Treatment in the Tumour Stage of Mycosis Fungoides Associated with the Appearance of Lesions in Organs other than the Skin

We have recently observed 3 patients with mycosis fungoides in the tumour stage, included in the programme of the Scandinavian Mycosis Fungoides Study Group, who responded satisfactorily to PUVA treatment yet developed signs of mycosis fungoides in other organs. These and similar findings (1, 6) lead us to believe the phenomenon to be important in the evaluation of effective topical remedies in this condition.

Total body painting with mechlorethamine (nitrogen mustard, HN2) solutions or megavolt electron beam radiotherapy or long-wave ultraviolet radiation after sensitization with a psoralen (PUVA) often lead to the disappearance of cutaneous lesions; and in the tumour stage of the disease PUVA treatment has recently been shown to produce fairly good results, the tumours often disappearing within one month (3).

As we shall demonstrate, satisfactory response to treatment or even apparent eradication of all traces of skin lesions does not necessarily prevent internal involvement or might even stimulate development of lesions in other organs. Three patients observed suffered from mycosis fungoides in the tumour stage. Three years earlier one had shown lymph-node involvement. At the time of starting PUVA treatment the disease was apparently confined to the skin in all three. All skin lesions cleared after 1–2 months of PUVA treatment.

Discussion

The rate of involvement of internal organs is far from well known, and completely contradictory reports have been made even by one and the same author. Rappaport & Thomas assume mycosis fungoides to originate in the skin, involvement of other organs being due to secondary spreading. They suggest on the basis of their necropsy findings (4) that a natural barrier exists in the skin itself, probably based on cell-mediated immunological mechanisms. The question arises as to whether PUVA therapy could interfere with a hypothetical mechanism of this nature: clinical results of treatment of conditions such as vitiligo, psoriasis and atopic dermatitis could theoretically be explained by similar mechanisms.

The natural history of mycosis fungoides is very

Table 1. The patients

<table>
<thead>
<tr>
<th>Year of Birth</th>
<th>Sex</th>
<th>Diagnosis</th>
<th>Starting PUVA</th>
<th>Non-skin lesions discovered</th>
<th>Sites other than skin</th>
<th>Previous treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1900</td>
<td>?</td>
<td>Trioxalen</td>
<td>April 1976</td>
<td>Whole of lower lip</td>
<td>X-ray</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>locally</td>
<td>July 1976</td>
<td>Intramuscular and</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>8-methoxalen</td>
<td>Oct. 1976</td>
<td>subcutaneous tumour</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>by mouth</td>
<td>July 1976</td>
<td>Kidney (at necropsy)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1902</td>
<td>?</td>
<td>Trioxalen</td>
<td>Aug. 1976</td>
<td>Inguinal node</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>locally</td>
<td>Jan. 1977</td>
<td>Base of tongue</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>8-methoxalen</td>
<td>March 1977</td>
<td>Subcutaneous tumour</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>by mouth</td>
<td>May 1977</td>
<td>Radiographic signs of</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>stomach involvement</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1915</td>
<td>?</td>
<td>Trioxalen</td>
<td>Jan. 1977</td>
<td>Left breast</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1966</td>
<td>?</td>
<td>locally</td>
<td>June 1977</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Acta Dermato-Venereologica (Stockholm) 58: 189-190, 1978
variable (2), and involvement of internal organs is open to question. We feel, however, that the rapid appearance of lesions at unusual sites shortly after commencing PUVA treatment of the patients presented is remarkable. Similar observations have also been made in three patients in conjunction with other topical therapy (1, 6), but as far as we are aware nothing like this has been noted in connection with systemic chemotherapy.

One important question remains to be answered. Could PUVA treatment have caused the appearance of mycosis fungoides lesions at these unexpected sites? If so, did true dissemination take place from a primary site in the skin, even when treatment seemed to have eliminated every trace of disease in this organ? One explanation would be the elimination of an immunologically operative barrier (see above). Alternatively, it is conceivable that the treatment destroys or neutralizes an antigen stimulus attracting the lymphoid cells to the skin (5).

If the phenomenon is not due to dissemination from a primary process in the skin, it is necessary to postulate a multifocal process, with microscopical, clinically undetectable changes, the growth of which might be stimulated in some unknown manner by the PUVA treatment. Here again, immunological interference—say with the much disputed surveillance mechanism—might be responsible.

Conclusion

Skin lesions of mycosis fungoides in the tumour stage can respond most dramatically to PUVA treatment. With its impressive immediate results, PUVA therapy is likely to become the treatment of choice for this disease. However, as it could be suspected that the treatment may provoke the development of similar lesions in other organs, we urge every clinician to watch for and report any similar observations. If it becomes obvious that this side effect of treatment is a reality it may well become necessary to combine all forms of topical treatment with systemic chemotherapy.

REFERENCES


Received October 7, 1977

Lars Molin, Marcus Skogh, Gunnar Volden

1 Department of Dermatology
University Hospital
Linköping
Sweden

2 Department of Dermatology
Rikshospitalet
Oslo
Norway