

Spiegler-Fendt Type Lymphocytoma Cutis: a Case Report of Two Patients Successfully Treated with Interferon Alpha-2b

Sir,

Lymphocytoma cutis of Spiegler-Fendt type is a rare chronic B lymphocyte proliferation affecting the head and the neck in particular. It is characterized by solitary or grouped nodules or plaques which are purple or red and may reach a diameter of 3–5 cm.

Although lymphocytoma cutis is a benign disorder, it often runs a protracted, even life-long course. Progression to malignant B cell lymphoma in a small percentage of patients is suggested by long-term follow-up of these patients. A therapy of proven value for lymphocytoma cutis is, however, lacking. We report here 2 cases of lymphocytoma cutis which were successfully treated with interferon alpha-2b. Both patients had previously received various unsuccessful therapies, but responded well to interferon.

CASE REPORTS

Patient 1

A previously healthy 39-year-old man presented with a 6-year history of reddish infiltrated nodules on his head. The first pathological examination of a biopsy 3 months after the initial presentation of the nodule showed lupus erythematosus discoides. The patient was treated with oxychlorine 300 mg/day for 3 months without response. A new biopsy taken 6 months later showed lymphocytoma cutis of Spiegler-Fendt type. The patient received phenoxymethylpenicillin 1 MIU q.i.d. for 2 weeks with no improvement. Borrelia antibodies were negative. Gradually the biopsied lesions healed and new biopsies were taken as a curative treatment. The largest lesion was excised by plastic surgery. Biopsies and excisions provided improvement for some time, but the patient developed new nodules. Most of the biopsies were examined by a pathologist. All except one (which showed atypical nuclei and increased mitoses, suggesting possible lymphoma) showed lymphocytoma cutis. For the one showing possible lymphoma the findings were discussed with clinicians, but because the clinical picture was silent, no radiotherapy was suggested. Subsequently, a new biopsy showed lymphocytoma cutis.

Six years after the onset of lymphocytoma cutis and after various



Fig. 1. Patient 1 before treatment with interferon alpha-2b, showing reddish, infiltrated nodules on the head.

unsuccessful treatments, interferon therapy was suggested by the oncologist. At that time the physical examination showed two violaceous 1.5-cm diameter tumid nodules on the left side of the head and some smaller nodules around the head (Fig. 1). There were no enlarged lymph nodes, and the liver and the spleen were normal. The complete blood count was normal.

The treatment was initiated with interferon alpha-2b 2.5 MIU subcutaneously 3 times a week. After 1 month of treatment some response was already seen: there were no new nodules and the size of the pre-existing nodules had decreased. The treatment was continued for 3 months and after that all the lesions had healed completely (Fig. 2).

At the start of the treatment the patient experienced slight fatigue and myalgia, but after 1 month of treatment all the side-effects had disappeared. A complete blood count was taken every month during the treatment, and mild leukocytopenia was observed (3.1×10^9) after 1 month of treatment, but by the next measurement it had returned to normal. Otherwise the blood count was normal.

The patient remained asymptomatic for 1 year and 11 months and was satisfied with the treatment. However, after that the patient developed a small nodule on his head. The biopsy again showed lymphocytoma cutis of Spiegler-Fendt. Because there was only one small nodule, interferon was not re-introduced. Instead, a punch biopsy was taken, because that had offered remission earlier. The nodule disappeared, but after 2 months a new small nodule had developed. A biopsy was taken. We are now waiting to see whether new lesions will develop. If so, interferon treatment might be re-introduced.

Patient 2

A 44-year-old man presented with a 4-year history of cutaneous nodules on his back and arm. Biopsy of one nodule taken by a private dermatologist showed lymphocytoma cutis of Spiegler-Fendt type. Borrelia antibodies were negative. The private dermatologist had followed up the nodules but did not give any treatment.

At the time the patient was admitted to the dermatology department he presented with a large violaceous infiltrate on his back and one on his left arm. His blood count was normal. No treatment was given for lymphocytoma cutis, but it was decided to follow up the lesions.

The patient was evaluated once a year, and 2 new biopsies showed lymphocytoma cutis. In between these check-ups the patient had visited a private GP who had prescribed oral azithromycin and corticosteroids. The lesions had cured but soon recurred.

Two years and 7 months later the patient was re-evaluated at the Department of Dermatology in between the scheduled visits because



Fig. 2. Patient 1 17 months after interferon treatment, showing complete resolution of lymphocytomas.

the nodules had enlarged notably. At that time the physical examination showed a violaceous infiltration on the patient's back and some new reddish nodules surrounded by annular erythema on his left shoulder. A new biopsy again showed lymphocytoma cutis. Complete blood count was normal. It was now decided to try interferon therapy.

The treatment was initiated with interferon alpha-2b 2.5 MIU subcutaneously 3 times a week, as with patient 1. After 1 month of treatment the nodules had become lighter in colour and decreased in size, after 2 months only small bluish plaques were seen on his back and left shoulder, and after 3 months all the lesions had healed completely.

At the beginning of the therapy the patient was always slightly febrile after the interferon injection. After 2 months of treatment he experienced some fatigue, but it disappeared by the next control. Complete blood count taken monthly during the treatment showed a mild leukocytopenia (2.9×10^9) after 2 months of treatment, but normalized in the following month.

However, after 6 months the patient contacted the dermatology department, because he had noticed 3 new nodules on his back and shoulder. The nodules were similar to the previous ones, but smaller. The biopsy showed lymphocytoma cutis. Because the patient was about to move to another country and because the lesions were small, interferon treatment was not considered. The lesions were treated with intralesional triamcinolone hexacetonide. They disappeared, but cutaneous atrophy developed. The treatment was continued with topical mometasone with occlusion, but the patient then moved abroad and was lost to follow-up.

DISCUSSION

Previous therapies for lymphocytoma cutis include penicillin, topical and intralesional steroids, argon laser, radiotherapy and cryosurgery. None of these have offered a valuable therapy in the long run. Intralesional corticosteroids have been of periodical help, but relapses are common. As far as we know, interferons have not been documented previously for the treatment of lymphocytoma cutis.

During the last few years, interferons have showed good responses in primary cutaneous T-cell lymphomas and mycosis fungoides (1–8). Lately they have also been shown to be effective in primary cutaneous B cell lymphomas. Zetone et al. (9) successfully treated a primary cutaneous B cell lymphoma with intralesional interferon alpha-2a. Intralesional interferon alpha-2a also offered a complete clearing of lesions in a *Borrelia burgdorferi*-associated primary cutaneous B cell lymphoma, as reported by Kutting et al. (10). Wollina (11) got a complete response of primary cutaneous T-cell-rich B cell lymphoma with perilesional interferon alpha-2a (11). However, the mechanism by which interferon induced remissions in these disorders remains unclear.

Because interferons seem to be effective in treating low-grade

malignant primary cutaneous B cell lymphomas, it is logical to postulate that they could also be effective in treating benign B cell infiltrates of the skin. We treated 2 patients with lymphocytoma cutis of Spiegler-Fendt type with subcutaneous interferon alpha-2b. Both patients responded well to the treatment. These cases suggest that interferon alpha-2b could provide an alternative for patients with Spiegler-Fendt type lymphocytoma cutis.

REFERENCES

1. McLaughlin P. The role of interferon in the therapy of malignant lymphoma. *Biomed Pharmacother* 1996; 50 (3–4): 140–148.
2. Olsen EA, Rosen ST, Vollmer RT, et al. Interferon alfa-2a in the treatment of cutaneous T-cell lymphoma. *J Am Acad Dermatol* 1989; 20: 395–407.
3. Vegna ML, Papa G, Defazio D, et al. Interferon alpha 2-a in cutaneous T-cell lymphoma. *Eur J Haematol* 1990; 45: (Suppl 52): 32–35.
4. Friedmann D, Wechsler J, Delfau M-H, et al. Primary cutaneous pleomorphic small T-cell lymphoma. *Arch Dermatol* 1995; 131: 1009–1015.
5. Ross C, Tingsgaard P, Jorgensen H, et al. Interferon treatment of cutaneous T-cell lymphoma. *Eur J Haematol* 1993; 51: 63–72.
6. Yagi H, Tokura Y, Furukawa F, et al. Th2 cytokine mRNA expression in primary cutaneous CD30-positive lymphoproliferative disorders: successful treatment with recombinant interferon-gamma. *J Invest Dermatol* 1996; 107: 827–832.
7. Wyss M, Dummer R, Dommann SN, et al. Lymphomatoid papulosis-treatment with recombinant interferon alfa-2a and etretinate. *Dermatology* 1995; 190: 288–291.
8. Watson A. Photochemotherapy for mycosis fungoides: current status. *Australas J Dermatol* 1997; 38 (1): 9–11.
9. Zetone T, Catimel G, Barbet N, et al. Complete remission of a primary cutaneous B cell lymphoma treated with intralesional recombinant interferon alpha-2a. *Eur J Cancer* 1994; 30A (2): 246–247.
10. Kutting B, Bonsmann G, Metze D, et al. *Borrelia burgdorferi*-associated primary cutaneous B cell lymphoma: complete clearing of skin lesions after antibiotic pulse therapy or intralesional injection of interferon alpha-2a. *J Am Acad Dermatol* 1997; 36 (2 pt2): 311–314.
11. Wollina U. Complete response of a primary cutaneous T-cell-rich B cell lymphoma treated with interferon alpha-2a. *J Cancer Res Clin Oncol* 1998; 124 (2): 127–129.

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