Follicular Mucinosis Developing into Cutaneous Lymphoma

Report of Two Cases and Review of Literature and 64 Cases in Japan

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Two cases of follicular mucinosis which developed into lymphoma are reported. The infiltrative atypical lymphocytes proved to be T cells, which were identified by monoclonal antibodies. The association of follicular mucinosis and lymphoma in Japan is estimated to be 9.4%. *Key words: Follicular mucinosis; Alopecia mucinosa; Malignant lymphoma; T cell lymphoma.* (Received June 14, 1983.)

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Follicular mucinosis was first described by Pinkus in 1957 under the name of alopecia mucinosa. This is an inflammatory disease of unknown etiology. Though the majority of cases follow an uneventful course, many cases of follicular mucinosis that may develop into lymphoma have been reported.

CASE REPORT

Case 1

A 37-year-old man was seen in November 1978 because of a 3-year history of generalized pigmented cutaneous lesions on the trunk and extremities. Cutaneous examination showed brown pigmented, infiltrated plaques involving the trunk and extremities. A biopsy specimen taken from a pigmented plaque showed typical follicular mucinosis. The patient had been treated with topical steroid ointments at another hospital. Three years later, the cutaneous lesions became increasingly infiltrated and tumors developed over the trunk and extremities. The patient was hospitalized in January 1982 with the diagnosis of malignant lymphoma. At this time various staged lesions such as erythema, infiltrating plaques and nodules were noted in addition to lymphadenopathies.

Histological examination of biopsied specimens showed dense infiltrates of large atypical lymphocytes (Fig. 1). At this time we reexamined retrospectively sections of the initial biopsied specimen, and a few atypical lymphocytes were sparsely present around the mucinous changes of the follicle (Fig. 2). Electron microscopy showed atypical lymphocytes with hyperconvoluted or cerebriform nuclei as well as clustered dense bodies. An inguinal lymph node biopsy specimen showed nonspecific dermatopathic lymphadenitis. To elucidate the nature of the atypical lymphocytes, immunohistochemical investigations were performed using monoclonal antibodies directed against T cells such as Leu-1, Leu-2a and Leu-3a and antibodies against IgM, IgG, and IgA. The surface of these atypical lymphocytes stained positive with only Leu-1, while negative results were obtained with the other antibodies. With these findings, this case was considered as a T-cell lymphoma. Further

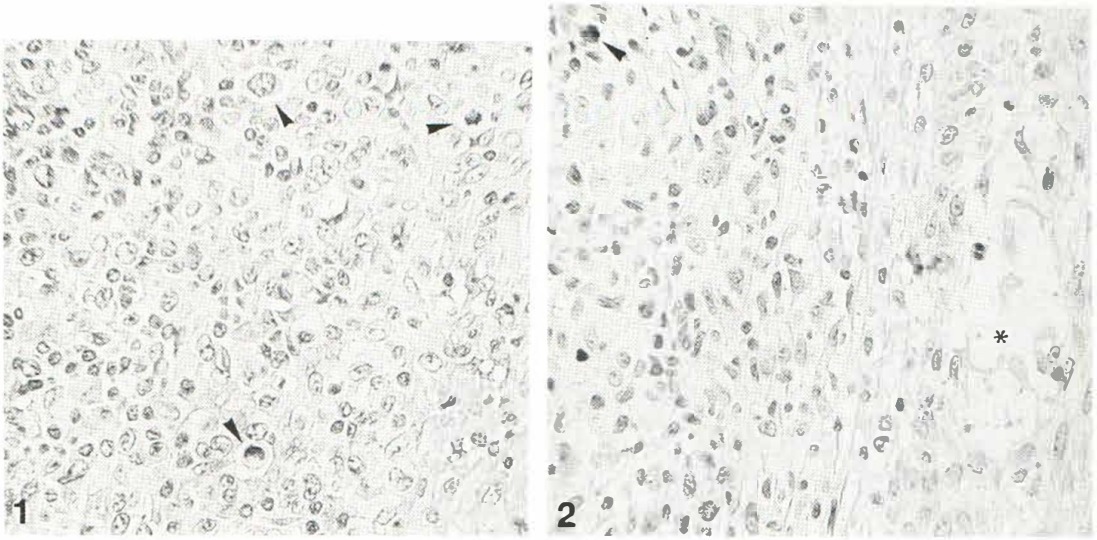

Fig. 1. Several atypical cells with enlarged cytoplasmas (arrow head). HE. ×200.

Fig. 2. An atypical cell (arrow head) seen in the infiltrative area around mucinous changed hair follicle (asterisk). HE. ×200.

lymphoma investigations, including complete blood counts, biochemical analyses, bone marrow biopsy, liver-spleen scan, lymphangiogram and systemic CT scan were negative. The patient was treated with total skin electron beam radiation twice weekly for 8 weeks. A total dose of 3 600 rad was given, using a six-field technique. At 2 200 rads the cutaneous lesions markedly improved leaving only pigmentation and few infiltrative atypical cells as seen histologically. Since then no recurrence has been noted for over a year.

Case 2

A 33-year-old man was first seen in May 1980. He had a nine-month history of localized erythematous pruritic lesions on the face, trunk and extremities. The lesion became slightly infiltrated and loss of hair was seen on the left eyebrow and the mustache. A biopsy specimen taken from infiltrating lesions was diagnosed as follicular mucinosis with no malignacy. The lesions failed to respond to topical steroid treatment and they persisted. In March 1982, the lesions became markedly infiltrated to form lots of plaques on the trunk and extremities. Systemic lymphadenopathies were also noted. A biopsy specimen taken from an infiltrated plaque revealed dense infiltrates of numerous pleomorphic atypical lymphocytes in the dermis in addition to the presence of Pautrier's microabscesses. At this time this case was diagnosed as Mycosis fungoides by a combination of clinical and histological findings. Electron microscopy showed atypical lymphocytes with hyperconvoluted nuclei. In vitro analysis of the atypical lymphocytes using monoclonal antibodies revealed that only Leu-1 and Leu-3a stained positively. From these findings, the parenchymal cells were suggestive of helper T cell (1). Further lymphoma investigations such as biochemical analyses, bone marrow biopsy and lymphangiogram were negative. This patient was also treated with total skin electron beam therapy (3 600 rads). Just after the therapy cutaneous lesions markedly improved both clinically and histologically. So far no recurrence has been noted for a year.

DISCUSSION

Follicular mucinosis is classified into three groups: localized benign, chronic benign and associated with a lymphoma (2). The last type, the association with a lymphoma was first reported by Braun-Falco in 1957 (3). Since then different authors reported cases of follicular mucinosis with lymphoma and analyzed the frequency of follicular mucinosis and

lymphoma as follows (4, 5, 6, 7, 8): 50% (5/10) (Kim et al. 1962), 13.3% (8/60) (Pinkus, 1964), 8.9% (8/90) (Plotnick et al., 1965), 66.7% (6/9) (Degos et al., 1966), 17% (8/47) (Emmerson, 1969) and 14% (7/50) (Coskey et al., 1970).

In Japan, sixty-four cases were collected and studied since 1960 (when the first case was reported in Japan) until 1982. With regard to the association of follicular mucinosis and lymphoma, six cases were reported to be associated, representing 9.4% of the total group. Therefore the association in the Japanese population seems to be fairly low as compared to data quoted in the literature except for Plotnick et al. (1965) who noted an even lower association. Four out of the 6 associated cases started out as follicular mucinosis and developed into lymphoma within 3 years. In the other two cases, one was reported as simultaneous occurrence of follicular mucinosis and lymphoma and the other was classified as a lymphoma first. Of the 6 cases 3 were in their thirties and 3 in their fifties. All but one case had generalized cutaneous involvement. This tendency of generalized cutaneous involvement was also referred to by Emmerson (1969) and Coskey et al. (1970). The ratio of male to female was 4 to 2. In analyzing the case studies, it was noted that in individuals beyond their thirties, having generalized cutaneous involvements and a chronic long course for more than 3 years, follicular mucinosis may be a forerunner of the malignant disease.

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