CNS Lymphoma Seen in Early Mycosis Fungoides

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MANCHESTER, ENGLAND — A 56-year-old man, who had experienced a skin eruption 6 years earlier and received a diagnosis of early-stage mycosis fungoides, which has a good prognosis, was hospitalized following a car accident and episodes of confusion.

He died 3 weeks later of a T-cell lymphoma involving the central nervous system, Dr. Aoife Lally said at the annual meeting of the British Association of Dermatologists.

Although there have been reports of CNS involvement in mycosis fungoides, it usually occurs in the setting of advanced and histologically transformed cutaneous disease. Isolated CNS involvement in stage 1B mycosis fungoides is “remarkably rare,” said Dr. Lally of the department of dermatology, Oxford (England) Radcliffe Hospitals.

When the patient originally was examined in the dermatology department, his skin findings included annular, erythematous, scaly patches and only a few plaques. There was no scalp or nail involvement and no lymphadenopathy.

Blood count, erythrocyte sedimentation rate, and renal and hepatic profiles were all normal.

The patient did not respond to treatment with potent topical steroids, and relapsed following a course of UVB treatment. Biopsies showed folliculotropic mycosis fungoides. Treatment with phototherapmy (PUVA) was commenced, and the patient responded well, according to Dr. Lally.

“At no point did he develop lymphadenopathy or systemic symptoms. But 5 years after initial presentation, a nodule appeared on the left ankle,” she said. This responded to radiotherapy, and the skin disease remained quiescent thereafter.

When he was admitted after the car accident, CT scan of the head revealed a frontal tumor. A craniotomy was performed, and histologic analysis confirmed the presence of a T-cell lymphoma.

“T-cell receptor gene analysis on DNA extracted from this tumor showed an identical T-cell clone to that of skin biopsies taken a year earlier,” Dr. Lally said.

Bone marrow examination was normal and staging CT found no nodal or extranodal disease.

The unusual nature of this disease course was echoed in a recent report of 10 cases of CNS involvement in mycosis fungoides, in the context of active skin disease, and with other visceral involvement (Cancer J. 2006;12:55-62). Stage at diagnosis also was significant: Only one patient had stage IB disease, with all others having stages 2B or greater. Mean survival following the onset of CNS symptoms in this cohort was approximately 5 months.

A second, recent report describes 10 patients with seemingly early stage mycosis fungoides that progressed to CNS involvement within just a few years.

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