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LETTER TO THE EDITOR

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A case of mycosis fungoides with small intestinal involvement

Dear Editor,

Mycosis fungoides (MF) is a malignant lymphoma derived from T cells. In most cases, MF presents only in the skin, although it may subsequently involve lymph nodes and other internal organs in advanced stages. Here, we report a case of MF with intestinal tract involvement.

A 67-year-old man presented with erythematous skin lesions on his arms, buttocks, and back that had existed for 3 years (Figure 1A, B). A skin biopsy from his back lesion showed epidermal infiltration of atypical lymphocytes (Figure 1C). The phenotype of infiltrating lymphocytes was $CD3^+$, $CD4^+$, $CD8^-$, $CD30^-$, and $CCR4^+$ (Figure 1D and data not shown). He was diagnosed as having plaque stage MF (T2bN0M0B0). He was treated with oral etretinate or etoposide in combination with narrow-band ultraviolet B therapy and occasional local electron beam irradiation. However, indurated tumors appeared on his trunk seven years after treatment with the drugs shown above (Figure 1E). Laboratory examination revealed elevated

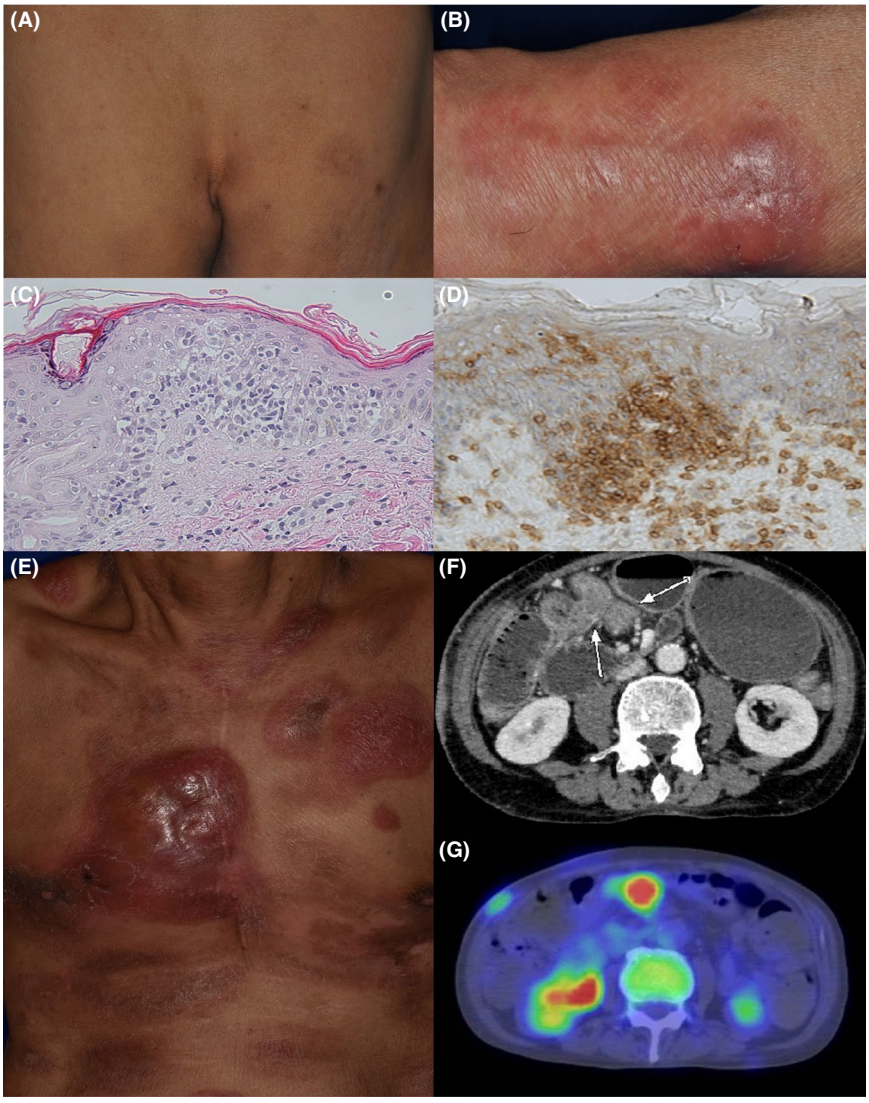

FIGURE 1 Erythema distributed on the buttocks (A) and the forearm (B). (C) Atypical lymphocyte infiltration into the epidermis (HE × 400). (D) Infiltrating lymphocytes positive for CD4 (×400). (E) Tumors on the trunk seven years after first examination. (F) Computed tomographic scanning showing small intestinal ileus. (G) Abnormal accumulation at the site of small intestines

serum levels of lactate dehydrogenase (314 IU/L), C-reactive protein (4.78 mg/dL), and soluble interleukin-2 receptor (sIL-2R; 2500 U/mL). As computed tomographic (CT) scanning and positron emission tomography (PET) found no organ involvement, mogamulizumab at a dose of 1 mg/kg was initiated, but it did not improve the skin lesions. Therefore, after four courses of mogamulizumab treatment, oral bexarotene at a dose of 300 mg/d was started. Eleven days after initiation of oral bexarotene, abdominal pain and vomiting were seen. CT scanning showed small intestinal obstruction (Figure 1F). Moreover, abnormal fluorodeoxyglucose accumulation with SUV max 9.4 was newly found by PET (Figure 1G). Serum sIL-2R level was increased up to 6800 U/mL and reflected the disease activity. As the disease activity was judged to be progressive, oral bexarotene was replaced by gemcitabine at a dose of 1000 mg/m^2. The skin lesions and small intestinal invasions responded to gemcitabine therapy; thereby serum sIL-2R level was decreased to 2500 U/mL after twice gemcitabine injections. However, the dose was reduced to 800 mg/m^2 because of pancytopenia, resulting in re-elevation at serum sIL-2R After totally five-time injections of gemcitabine, the patient died of a small intestinal perforation.

As far as we know, there have been seven case reports of MF with intestinal involvement.[1–4] All the cases had death outcomes in less than a year after occurrence of intestinal involvement. Regardless of various treatment including surgical operations and multi-agent chemotherapy, mean survival time was three months.[1–4] Consistently, this patient died 63 days after intestinal involvement happened. Considering the fact that median survival time of stage IVB MF was 33 months,[5] intestinal involvement in advanced stage MF represents extremely poor prognosis.

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CONFLICT OF INTEREST

The authors declare no conflict of interest.

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REFERENCES

1. David NS, Stanley SB, Stephen B. Gastrointestinal complications of mycosis fungoides. J Royal Soc Med. 1984;77:114–9.
2. Chen K-R, Tanaka M, Miyakawa S. Granulomatous mycosis fungoides with small intestinal involvement and a fatal outcome. Br J Dermatol. 1998;138:522–5.
3. Poonam V, Mohit T, Ike U, David G. Small bowel obstruction due to mycosis fungoides: an unusual presentation. Am J Med Sci. 2011;341:508–9.
4. Álvaro A, Rómulo D. Unusual involvement in mycosis fungoides: duodenal papilla. Rev Esp Enferm Dig. 2016;108:513–6.
5. Scarisbrick JJ, Prince HM, Vermeer MH, Quaglino P, Horwitz S, Porcu P, et al. Cutaneous lymphoma international consortium study of outcome in advanced stages of mycosis fungoides and sézary syndrome: effect of specific prognostic markers on survival and development of a prognostic model. J Clin Oncol. 2015;33:3766–73.