Abstract

Paraneoplastic pemphigus is a rare autoimmune blistering disease generally associated with malignancy. The clinical presentation consists typically of painful and diffuse erosive stomatitis that may be accompanied by polymorphic skin lesions and systemic involvement. Diagnosis is based on clinical manifestations and confirmed by histology and immunological testing. The current first-line treatment is systemic corticosteroids and adjuvant therapies, including immunosuppressive agents. We report a case of buccal paraneoplastic pemphigus resistant to ibrutinib and rituximab successfully treated with azathioprine and polyclonal immunoglobulins.

Keywords: Autoimmune blistering disease; Chronic lymphocytic leukemia; Paraneoplastic autoimmune multiorgan disease; Paraneoplastic pemphigus.