[PICTURES IN CLINICAL MEDICINE]

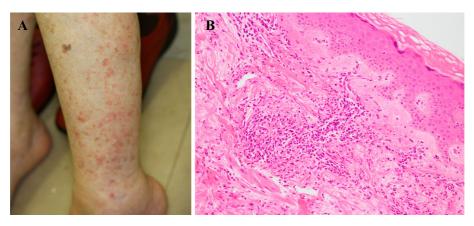
Schnitzler Syndrome after COVID-19 Vaccination

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Key words: Schnitzler syndrome, COVID-19 vaccination, lymphoproliferative disorders

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Picture.

An 86-year-old Japanese woman was referred for an examination of leukocytosis and diagnosed with Waldenström macroglobulinemia. She was asymptomatic and remained stable during follow-up without treatment. The day after receiving her first COVID-19 vaccination (Pfizer-BioNTech), she developed urticarial exanthema on her limbs. Although the rash improved with topical steroids, it worsened the day after she received the second vaccination; bone pain, general fatigue, and leukocytosis (white blood cell count: 11,600/ μL) also developed. A skin biopsy showed perivascular and interstitial neutrophil infiltrate with leukocytoclasia (Picture A, B), and definite Schnitzler syndrome was diagnosed according to the Strasbourg criteria (1). She rapidly responded to low-dose prednisolone (10 mg/day), and the skin rash and systemic symptoms resolved. Schnitzler syndrome is autoinflammatory and characterized by urticarial rash and immunoglobulin M monoclonal gammopathy. Lymphoproliferative disorders feature immune dysregulation that often results in autoimmune disease (2). In patients with lymphoproliferative disorders, physicians should pay careful attention to immune complications after COVID-19 vaccination.

The authors state that they have no Conflict of Interest (COI).

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