Extranodal natural killer/T-cell (ENK/T) lymphoma is a rare neoplasm, subcategorized into ENK/T-nasal (ENK/T-N) and ENK/T-nasal type (ENK/T-NT) lymphomas. ENK/T-NT lymphoma with initial presentation in the skin is known as primary cutaneous (PC) ENK/T-NT lymphoma.

Patients and methods: The aim of this study was to investigate pathogenesis, genomic alterations, and prognosis of cutaneous ENK/T lymphomas to provide further insights into clinicopathologic features and genetic mechanism of lymphomagenesis. A retrospective case study of 5 white patients affected by ENK/T lymphoma (4 PC-ENK/T-NT and 1 ENK/T-N with cutaneous involvement) was performed.

Results: Most of the cases presented with multiple nodular and ulcerated lesions localized on the extremities. A considerable percentage had disease in advanced stage at diagnosis with a 12-month survival rate of 40%. Genomic alterations were detected by array-based comparative genomic hybridization that showed gains of 1q, 7q and loss of 17p in the cases of PC-ENK/T-NT lymphomas and gain of 7q and loss of 9p, 12p, 12q in the case of ENK/T-N lymphoma. Conclusion: ENK/T lymphoma is a very aggressive entity, and, in our cases, the exclusively cutaneous presentation was not associated with a better prognosis. The results of our array comparative genomic hybridization analysis could be useful to better define the different ENK/T lymphoma subgroups with cutaneous involvement and new protocols of treatment.