

Paraneoplastic pemphigus in Russian patients: A single center case series



Abstract

Paraneoplastic pemphigus (PNP) is a rare autoimmune mucocutaneous disease with mortality rates up to 90%. Here, we retrospectively provide the first description of a PNP case series in the Russian population seen at our department over a 7-year period. In our study, PNP and neoplasm occurred among patients aged >40 years. Seven patients had cancer of different localization, and one patient suffered from chronic lymphocytic leukemia. The duration of malignancy ranged from 1.1 to 7 years (mean: 4 years). Pemphigus was diagnosed before and after malignancy in two (Table 1, cases #2 and 5) and six patients, respectively. Surgical interventions were performed in six cases, and chemotherapy was used in three patients. These treatment modalities were successful in six patients (improvement in five cases and stabilization of leukemia in one patient). Three of eight patients (37.5%) died during the follow-up period as a result of metastases and/or multiorgan failure. We assumed that overall prognosis of PNP depends on the type and the stage of the associated tumor. Lung cancer is characterized by one of the highest mortality rates (the overall 5-year survival rate is 17.7–55.2%) among all types of cancer. In all patients who survived, the treatment of both PNP and malignancy was effective, suggesting that effective treatment of cancer (especially tumor resection) could improve the prognosis of PNP. On the other hand, in the case of an unresectable tumor, treatment of the underlying malignancy is often not associated with the improvement of PNP. Thus, future research is needed to reveal factors and features of PNP that must be taken into account when determining the disease prognosis and treatment strategy.

Publications

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Biography

Anfisa Lepekhova has her expertise in severe cutaneous drug reactions and autoimmune blistering disorders. Research lines of the past 5 years: genetic mechanisms in autoimmune bullous dermatoses, investigation of blister fluid biomarkers in severe cutaneous drug reactions; non-scarring alopecia treatment. Doctor is the head of the educational process at the department of skin and venereal diseases (Sechenov University).