



AUTOIMMUNE BULLOUS DISEASES

IMMUNOFLUORESCENCE CYTOLOGY IN THE DIAGNOSIS OF OCULAR PEMPHIGUS VULGARIS

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Background: Pemphigus vulgaris (PV) is a mucocutaneous autoimmune bullous disorder. The hallmark of pemphigus is acantholysis, the loss of cell-cell adhesion. Ocular involvement is rare, and nonscarring conjunctivitis and erosions are the main findings. Atypical presentations may require immunopathologic assays, which are not frequent available. Conjunctival biopsy is, however, a difficult and uncomfortable procedure. Tzanck's smear is a sensitive diagnostic tool for PV, but cytomorphology of acantholytic cells may mimic malignancy, as previously described in oral and vulvovaginal pemphigus. Immunofluorescence cytology increases its specificity, demonstrating the relevance of a fast and little invasive technique for the diagnosis of atypical forms of PV.

Observation: A 51-year-old female presented with erosions on skin and variable involvement of oral, ocular, vaginal, perianal and esophageal mucosae since 2005. Diagnosis of PV was confirmed by immunopathological examination of skin and cervix fragments. Treatment included topical corticosteroids, oral prednisone and immunosuppressive drugs, such as dapsone, azathioprine and mycophenolate mofetil. The clinical course was refractory, despite all combined therapies. In 2014, she developed an erythematous lesion with atypical vascular pattern on the nasal conjunctiva of the right eye. Conjunctival cytology demonstrated atypical cells and supported its surgical removal due to suspicion of neoplasm. Postoperative histology detected intraepidermal acantholysis and immunohistochemistry revealed intercellular intraepidermal IgG4 deposits, confirming the diagnosis of PV. Topical antibiotics and corticosteroids promoted transitory improvement. In 2016, the patient started with bilateral blepharitis and conjunctival ulcers. Immunofluorescence cytology of ocular smear showed intercellular IgG deposits around acantholytic cells, confirming ocular PV and discarding neoplastic aggression.

Key message: To our knowledge, this is the first report to illustrate misinterpretation of acantholytic cells on conjunctival cytology. Immunofluorescence cytology increase its specificity and provides the diagnosis of ocular pemphigus.

