Disclosure page:

No disclosures

A case of progressive immunotherapy-related mucocutaneous eruption (PIRME) in the setting of ipilimumab/nivolumab for metastatic squamous cell carcinoma

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A 34-year-old male with metastatic penile squamous cell carcinoma presented with painful erosions, hemorrhagic crusting, and bullae involving his oral mucosa, sacrum, and left lower abdomen. The patient had completed his second cycle of ipilimumab/nivolumab five days prior to onset. Laboratory workup was remarkable for anemia and thrombocytosis. Respiratory pathogen panel, throat culture for Group A streptococcus, and HSV/VZV swabs were negative. Punch biopsy demonstrated vacuolar alteration of the basal cell layer with separation at the dermal epidermal junction and overlying full-thickness epidermal necrosis. Oral prednisone was initiated at 0.5 mg/kg/day dosing and led to significant clinical improvement. Given milder course with rapid improvement with oral steroids, lack of ocular involvement, and onset in the setting of immunotherapy, the diagnosis of progressive immunotherapy-related mucocutaneous eruption (PIRME) was made. PIRME is a recently described clinical entity of bullous mucocutaneous eruptions that are clinically and pathologically similar to Stevens-Johnson syndrome/toxic epidermal necrolysis (SJS/TEN) but occur in the setting of recent immunotherapy and typically follow a more indolent course. Ocular involvement is rare compared to SJS/TEN. In PIRME, some cases are thought to be triggered by a "second-hit" medication following priming of the immune system with initiation of immunotherapy, although in this case, we were unable to definitively identify a second culprit medication. Given progression of our patient's metastatic disease, the decision was made to re-trial the patient on ipilimumab/nivolumab due to high suspicion for PIRME. He successfully restarted treatment with ipilimumab/nivolumab without recurrence of mucocutaneous symptoms and is currently tolerating his seventh cycle.

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