Eosinophilic granulomatosis with polyangiitis exacerbated by dupilumab

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Case presentation

A 60-year-old male presented to the emergency department with worsening shortness of breath, cough with blood tinged-sputum, headache, fever, and rash. Twenty months prior, he developed asthma-like symptoms which resulted in several hospital admissions for acute hypoxemic respiratory failure. Each exacerbation improved modestly with short prednisone tapers, but respiratory symptoms persisted between admissions, eventually leading to a diagnosis of occupational asthma. Ten days prior to presentation, he initiated dupilumab.

At presentation, he had an eosinophilia of 3400 and pulmonary infiltrates on imaging. Scattered on the upper and mid back, there were palpable violaceous papules and plaques. Given new onset of rash with a question of temporal relationship to dupilumab, biopsy was performed. Leukocytoclastic vasculitis with an abundance of perivascular eosinophils on histopathology led to further workup of P-ANCA positivity and a diagnosis of eosinophilic granulomatosis with polyangiitis (EGPA). He improved on high-dose methylprednisolone, with eventual taper and plans to transition to mepolizumab, an IL-5 inhibitor. The initiation of dupilumab prior to symptom progression raised the question of whether dupilumab precipitated a vasculitic phase.

Acknowledgements

We thank the patient for agreeing to let us share his case.

Figure 1. Skin biopsy revealing leukocytocvlastic vasculitis with an abundance of perivascular eosinophils

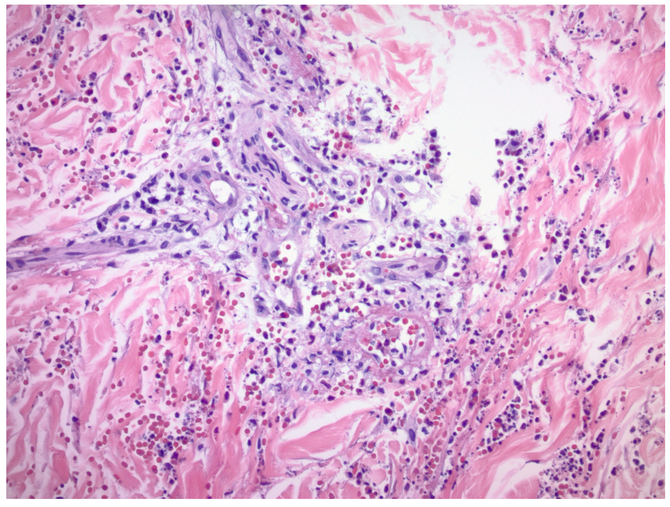
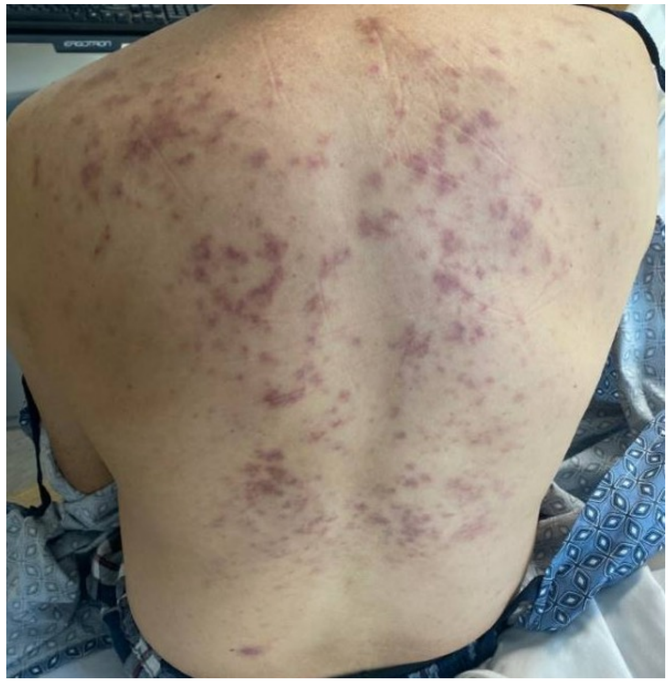


Figure 2. Patient skin eruption upon presentation to emergency department, with purpuric plaques localized to the back



Discussion

- There have been several cases in the literature of dupilumab exacerbating or "unmasking" EGPA, as well as 6 cases of EGPA reported as adverse events in dupilumab clinical trials for asthma and chronic rhinosinusitis with nasal polyps[1,3-8].
- The potential mechanism by which dupilumab may unmask EGPA is through uninhibited IL-5 induced eosinophil activity in the periphery[2].
- To our knowledge, there have been no cases of EGPA reported for patients receiving dupilumab specifically for atopic dermatitis.
- Dermatologists should be cognizant of dupilumab's potential to reveal or aggravate conditions characterized by eosinophilia.

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