doi:10.36849/JDD.2020.164

Remission of Refractory PASH Syndrome Using Ixekizumab and Doxycycline

Muhammad I. Gul BS,[a] Vivek Singam BLA,[b] Cody Hanson MD,[c] Brett C. Neill MD,[c] Daniel J. Aires MD JD,[c] Anand N. Rajpara MD,[c]

[a]University of Kansas School of Medicine, Kansas City, KS
[b]University of Missouri-Kansas City School of Medicine, Kansas City, MO
[c]University of Kansas Medical Center, Kansas City, KS

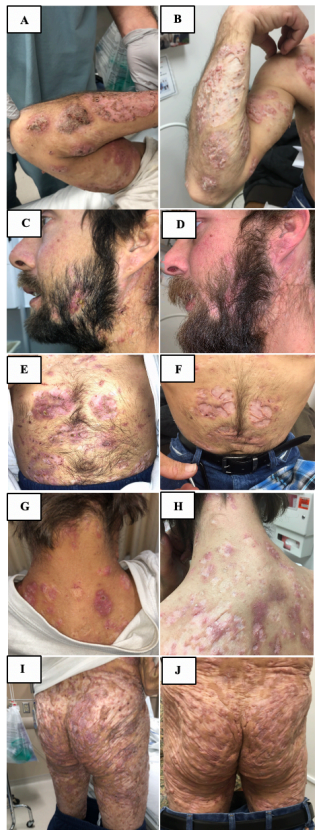
Ixekizumab is a relatively new IL-17 blocking agent recently approved for treatment of psoriasis. We present a first report of successful treatment of PASH syndrome using ixekizumab (Taltz®) and doxycycline in a patient who failed other therapies.

A 37-year-old Caucasian male with a familial history of PASH syndrome presented to the clinic for fatigue, pain and worsening of his cutaneous lesions. At presentation the patient had a 10-year history of vegetative pyoderma gangrenosum (PG), acne fulminans, and Hurley Stage IV hidradenitis suppurativa (HS). His condition was refractory to a 2-month trial of clindamycin and Rifampin PTA therapy, a 4-month trial of adalimumab, and a 5-month trial of ustekinumab. Since Cugno et al recently noted that IL-17 is overexpressed in PASH skin lesions,1 we started the patient on ixekizumab (Taltz®) therapy with a 160 mg loading dose, followed by 80 mg maintenance doses every 2 weeks. Doxycycline 100 mg twice daily was started at the same time. 12 weeks after starting these treatments his PG, acne conglobate and HS remitted (Figure 1). The patient reported significant improvement in quality of life with decreased fatigue and pain.

While PASH has historically have been managed with PTA therapy and surgery, these have shown limited success.1 Biologic agents such as adalimumab, anakinra, and ustekinumab have recently demonstrated safety and efficacy for certain but not all PASH patients.1 Although recent studies have revealed IL-17 plays a critical role in the pathogenesis of PASH,2 to date, there are no reports regarding efficacy of the IL-17-blocker ixekizumab in PASH syndrome.

In December 2017, the FDA approved ixekizumab for treatment of plaque psoriasis and psoriatic arthritis. Compared with other biologic agents such as TNF-blockers, ixekizumab may prove more effective in management of PASH patients due to its IL-17 blocking activity since IL-17 drives several key steps in PASH pathogenesis.[1] These findings may have important implications in future management of PASH patients, especially those who do not respond to PTA therapy, surgical intervention or to other biologic agents.

FIGURE 1. Patient at the time of presentation (left) and after 12-week therapy with ixekizumab (Taltz®) and doxycycline (right).



DISCLOSURES

The authors have no conflicts of interest to disclose.

References

1. Cugno M, et al. "PAPA, PASH and PAPASH syndromes: Pathophysiology, presentation and treatment." *Am J Clin Dermatol.* 2017 Aug;18(4):555-562.
2. Marzano AV, Ceccherini I, Gattorno M, Fanoni D, Caroli F, Rusmini M, et al. Association of pyoderma gangrenosum, acne, and suppurative hidradenitis (PASH) shares genetic and cytokine profiles with other autoinflammatory diseases. *Medicine (Baltimore).* 2014;93(27):e187

AUTHOR CORRESPONDENCE

Anand Rajpara MD
E-mail:..... arajpara@kumc.edu