Comparison of Generalized Pustular Psoriasis Patients With and Without Documented Flare Episodes in Their Medical Record

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BACKGROUND & STUDY OBJECTIVES

- Generalized pustular psoriasis (GPP) is a rare, but severe multisystem, chronic inflammatory disease characterized by sudden and widespread eruption of sterile pustules of varying severity.[1-3]
- Pustules during and after a GPP flare can often last for many weeks and can lead to life-threatening complications often requiring emergency care.[1-4]
- The most recent estimate of mortality is 4.2% from a nationwide study of hospitalized patients with GPP in Japan.[5]
- There is growing evidence focused on understanding the overall disease burden and treatment patterns in patients with GPP; however, there is minimal research documenting characteristics of GPP flares.[4,6]
- The objective of this study is to describe differences between patients with GPP who do (GPP flare) and who do not (GPP no flare) have flare episodes documented in their electronic health record (EHR).

METHODS

- This retrospective descriptive study included adult patients with GPP (ICD-10 code L40.1) identified in the Optum® de-identified EHR data between 1 July 2015-30 June 2020.
- The index GPP diagnosis was the first occurrence in the EHR with no coded history of GPP for at least one year prior.
- Only patients with at least 12 months of health care activity documented in the EHR after the index diagnosis and with notes available in the EHR were included in the study.
- EHR "terms" refer to signs, diseases, and symptoms (SDS) documented, and "attributes" are descriptions of the SDS terms. Categories were developed to capture like words of the same concept. Each health care visit beginning on or after the index date of GPP diagnosis with documented notes that met at least one of the following criteria were considered a documented flare in the EHR.
  1. For visits with a primary diagnosis of GPP (ICD-10 code L40.1):
     a. An encounter with place of service = 'EMERGENCY PATIENT', 'OBSERVATION PATIENT', 'INPATIENT', or 'URGENT'.
     b. Any term in the "Flare" category
     c. Any term in the "Pustule/Lesion" category plus any flare attribute
     d. Any term in the "Rash" category plus any flare attribute
     e. Any term in the "Other GPP symptoms" category plus any flare attribute
  2. For dermatology-related visits (i.e., type of provider was a dermatologist or primary reason for visit was any skin-related diagnosis defined by ICD-10 L* or R2* code):
     a. Any term in the "Flare" category AND any term in the "Pustule/Lesion" category
     b. Any term in the "Pustule/Lesion" category plus any flare attribute
- Flare episodes were defined as consecutive days that a flare was documented in the EHR and were characterized by the frequency of occurrence per patient, the setting of care where they were identified, the type of specialist managing the episode, associated symptoms, and the treatments before, during, and after the episode.

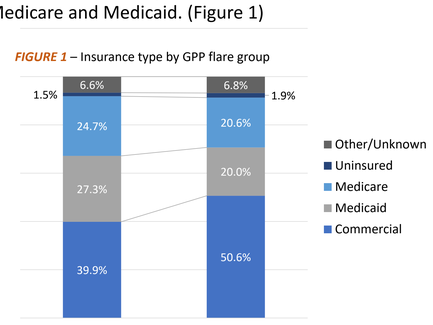
RESULTS

- Of the 48.6 million patients with EHR notes available, 1,535 patients with GPP were identified, and 271 of those patients had at least one flare episode documented in their EHR, accounting for a total of 513 flare episodes during the study period.
- Age and gender were similar among GPP flare and GPP no flare patients (Table 1), but flare patients were more likely to be enrolled in Medicare and Medicaid. (Figure 1)

TABLE 1 – Demographics of patients with GPP

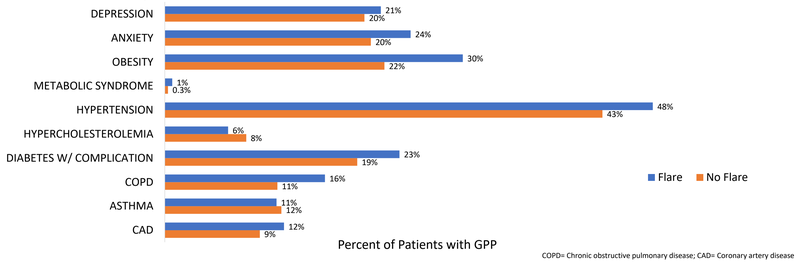
	Total patients	GPP flare patients	GPP no flare patients
Age, *mean [SD]*	53.4 [14.7]	53.5 [15.2]	53.3 [14.5]
Gender, *% female*	66.3%	66.8%	66.2%

FIGURE 1 – Insurance type by GPP flare group



- Patients with documented flares had a 24% higher score on the Charlson Comorbidity Index and were more likely to be obese (30% vs 22%), have hypertension (48% vs 43%), anxiety (24% vs 20%), diabetes with complications (23% vs 19%), COPD (16% vs 11%), and coronary artery disease (12% vs 9%). (Figure 2)
- Few to no differences between the groups were found with depression (21% vs 20%), hypercholesterolemia (6% vs 8%), and asthma (11% vs 12%). (Figure 2)

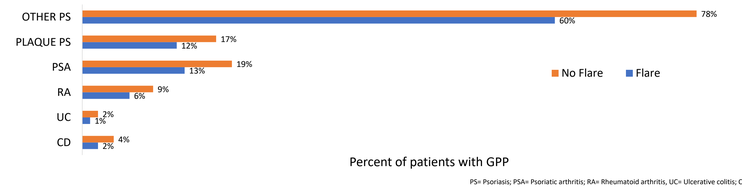
FIGURE 2 – Comorbidity of Patients with GPP



COPD= Chronic obstructive pulmonary disease; CAD= Coronary artery disease

- GPP flare patients were also more likely to have concurrent autoimmune conditions such as plaque psoriasis (17% vs 12%), psoriatic arthritis (19% vs 13%), and rheumatoid arthritis (9% vs 6%). (Figure 3)

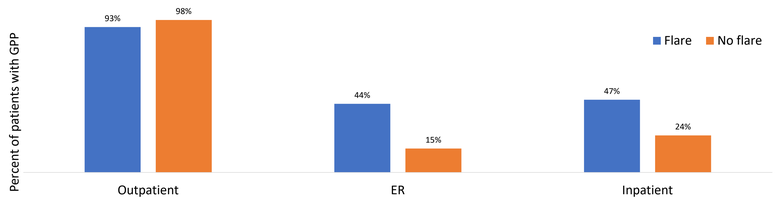
FIGURE 3 – Autoimmune conditions of Patients with GPP



PS= Psoriasis; PSA= Psoriatic arthritis; RA= Rheumatoid arthritis, UC= Ulcerative colitis; CD= Crohn's disease

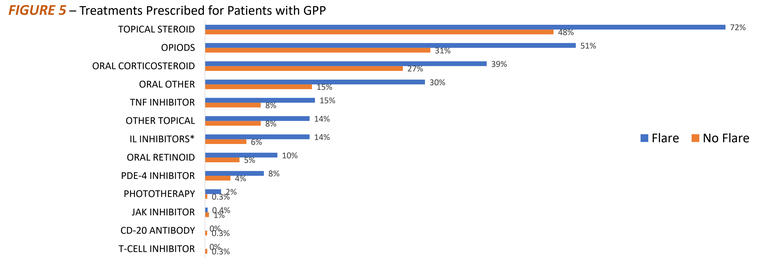
- Although little to no differences between the groups in outpatient visits, GPP flare patients were almost three times more likely to have any ER visits (44% vs 15%) and almost twice as likely to have any inpatient visits (47% vs 24%). (Figure 4)
- The average length of any hospital stay was 4 days for patients with documented flares and 3 days for those who did not have documented flares.

FIGURE 4 – Utilization of Any Inpatient and ER Services Among Patients with GPP



- GPP flare patients had greater use of drug treatments across almost all classes, including topical steroids (72% vs 48%), oral corticosteroids (39% vs 27%), other oral dermatologic medications (30% vs 15%), TNF inhibitors (15% vs 8%), other topicals (14% vs 8%), and IL-17, IL12/23 or IL23 inhibitors (14% vs 6%). (Figure 5)
- 51% of flare patients were prescribed opioids compared to 31% of no flare patients. (Figure 5)

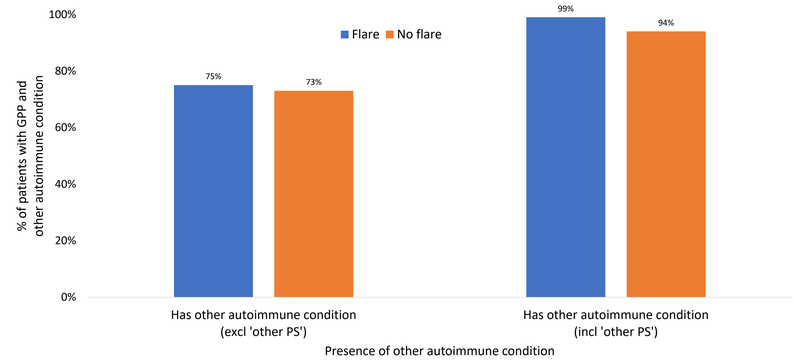
FIGURE 5 – Treatments Prescribed for Patients with GPP



* IL INHIBITORS include IL-17, IL12/23 and IL23 inhibitors

- Regardless of whether patients flare or not, biologic use is driven by other autoimmune conditions. (Figure 6)

FIGURE 6 – Presence of Other Autoimmune Conditions Among Those Treated with Biologics



CONCLUSIONS

- Patients with GPP and documented flares had a greater disease burden than those who did not have documented flares in their EHR.
  - Flare patients had more co-morbidities than non-flare patients.
  - Flare patients experienced higher rates of ER and inpatient visits.
- There is a significant, unmet need for the treatment of flares in patients with GPP.
  - Advanced treatment use among patients with GPP was low and may have to be dictated by the presence of other autoimmune conditions.
  - The use of topical steroids was higher among patients with GPP and documented flares than those that did not have documented flares in their EHR.
- More frequent use of opioids suggests that pain is a significant burden for patients with GPP and documented flares.

LIMITATIONS

- Due to the rarity and lack of awareness of GPP, it is likely that additional patients with true GPP remain undiagnosed or are miscoded with another form of psoriasis. In order to ensure that the study captured patients with GPP, only patients coded with a GPP diagnosis were included.
- The algorithm to identify GPP flares was intentionally conservatively designed to ensure that only flares due to GPP were identified. Thus, it is likely that the methodology underestimates the number of GPP episodes.
- Patient surveys suggest that patients do not always seek medical treatment for flares. This study only identifies patients with GPP flares documented in their EHR and underestimates the true number of patients with GPP who flare and the number of flare episodes. Also, documented flare episodes are likely to be higher severity than those not documented.
- In the EHR data, the treatments are based on prescriptions written or administered in the office/facility. Written prescriptions may not represent whether the prescription was filled and/or actually taken by the patient.
- Duration of flare episodes cannot be accurately determined in EHR data.

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