Correlation of early-onset hidradenitis suppurativa with stronger genetic susceptibility and more widespread involvement

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Background: The reported mean age of onset of hidradenitis suppurativa (HS) is between 20 and 24 years. Prepubertal onset is thought to be rare.

Objective: We sought to determine the prevalence of early-onset HS and to compare clinical characteristics between early-onset and normal-onset HS in a retrospective study.

Methods: Data were collected from 855 patients with HS. Early-onset HS was defined as onset before the thirteenth birthday. Clinical characteristics were analyzed in relation to the age of onset.

Results: In all, 66 patients (7.7%) reported early-onset HS. A family history for HS was significantly higher in early-onset patients (55.6% vs 34.2%; odds ratio 2.1, 95% confidence interval 1.2-3.6, $P = .006$). They developed inflammatory lesions at more body sites than patients with normal-onset HS (odds ratio 3.0, 95% confidence interval 1.8-4.9, $P < .001$). Distribution of the Hurley stages of severity showed no differences between the 2 groups (odds ratio 1.1, 95% confidence interval 0.7-1.8, $P = .72$).

Limitations: Some data were based on patient-reported information.

Conclusion: Early-onset HS occurs more frequently than previously believed. Patients with early-onset HS often report a family history for HS and develop lesions at more body sites. (J Am Acad Dermatol 2015;72:485-8.)

Key words: acne ectopica; acne inversa; age of onset; disease extent; disease severity; early onset; family history; genetic predisposition; hidradenitis suppurativa.

Hidradenitis suppurativa (HS) is characterized by recurrent inflammatory nodules at intertriginous body sites.[1,2] Onset of HS is generally after puberty, typically between the ages of 20 and 24 years.[3-5] Prepubertal onset is estimated to occur in 2% of patients with HS.[6,7] When several of our patients reported onset of HS in their early teens, we sought to determine the prevalence of early-onset HS and to analyze the clinical characteristics of patients with HS in relation to the age of onset.

METHODS

Patients

We collected data from 855 patients given the diagnosis of HS between 2007 and 2014 in The Netherlands at the Department of Dermatology at the Erasmus University Medical Center, Rotterdam; the Deventer Hospital, Deventer; and the Department of Plastic Surgery at the Diaconessenhuis, Leiden. Data extraction took place partly from our HS database, also used by Schrader et al.[5] For this type of retrospective analysis, no

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medical ethical committee approval is required under Dutch law.

Data collection

Patient characteristics were collected from the medical files. A family history was considered positive when a first- or second-degree relative had HS. The Hurley classification was used to assess disease severity.[2] Body sites counted were: axillary, inguinal/genital, perianal, gluteal, abdominal, and (infra) mammary. Normal weight was defined as a body mass index (BMI) below 25 kg/m^2, overweight as a BMI between 25 and 30 kg/m^2, and patients with a BMI above 30 kg/m^2 were categorized as obese.

CAPSULE SUMMARY

- Prepubertal onset of hidradenitis suppurativa is believed to be rare.
- Our study found early-onset hidradenitis suppurativa to be more common than anticipated, with a stronger genetic predisposition and more widespread disease at a later age.
- Patients with early-onset hidradenitis suppurativa should be closely monitored to receive early and adequate treatment.

Early- and normal-onset HS

Early-onset HS was defined as reported onset of disease before the thirteenth birthday. This cutoff point was chosen based on the mean age at menarche of 13.5 years in The Netherlands.[8] In general, puberty occurs later in boys. Therefore, it is conceivable that most patients with onset before their thirteenth birthday were prepubertal or in early puberty.

Data analysis

Statistical analysis was performed using SPSS Statistics 21 (IBM Corp, Armonk, NY). Independent *t* tests were performed for nominal data, presented as mean ± SD. Pearson χ^2 test was used for categorical data, and presented as number (%). Binary logistic regression was used to investigate the effect of family history on early-onset HS, corrected for age and gender. As a second step we took early onset as a predictor for disease severity, ie, Hurley stage and number of locations affected. Here the response variables were ordinal and we used ordinal logistic regression, correcting for age and gender. *P* values less than .05 were considered significantly different.

RESULTS

A total of 620 women and 235 men, mean age 37.9 ± 12.8 years, were included (Table I). Body sites most frequently affected were the inguinal/genital (89.7%), axillary (64.3%), and gluteal (41.2%) areas. The (infra) mammary (20.7%), perianal (19.5%), and abdominal (16.7%) regions were least affected. Most patients were current smokers (70.5%) or ex-smokers (14.2%), who started smoking at a mean age of 16.1 ± 3.9 years.

Early-onset associated with a positive family history for HS and more widespread disease

Early onset was reported by 66 patients (7.7%) and was related to a significantly higher percentage reporting a family history of HS (55.6% vs 34.2%, P = .001); this remained significant after correcting for age and gender (odds ratio 2.1, 95% confidence interval 1.2-3.6, P = .006). Early onset was associated with significantly more affected body sites (Table I). This association remained significant after correcting for age and gender (odds ratio 3.0, 95% confidence interval 1.8-4.9, $P < .001$). No difference was found in the distribution of Hurley stages, even after correcting for age and gender (odds ratio 1.1, 95% confidence interval 0.7-1.8, P = .72).

Fewer early-onset patients were smokers whereas there was no difference in the BMI

Of the 696 current smokers and ex-smokers, only a few of the early-onset group reported smoking before the onset of HS, whereas most of the normal-onset group had smoked before they developed symptoms (3/46, 6.5% vs 568/650, 87.4%, $P < .001$). There was no difference in weight in the BMI subgroups (Table I), nor in mean BMI (28.6 ± 7.0 vs 28.0 ± 6.0, P = .48) between early-onset and normal-onset HS.

No differences in comorbidities between early-onset and normal-onset HS

No difference was found among the percentage of patients reporting acne during puberty (15/56, 26.8% vs 146/580, 25.2%, P = .12), concomitant rheumatoid arthritis (3/61, 4.9% vs 38/645, 5.9%, P = .09) or inflammatory bowel disease (1/66, 1.5% vs 24/784, 3.1%, P = .63).

DISCUSSION

In this retrospective study of 855 patients, 66 (7.7%) reported early-onset HS. This is higher than previously reported (2%),[7] indicating that early-onset HS is not so rare. Our results suggest that for HS, as with psoriasis and atopic dermatitis,[9,10]