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The American Journal of Kidney Diseases

ISSN 3064-6642

Case Report

Newly Diagnosed FSGS And Vasculitis In A Patient, Known Case Of Hidradenitis Suppurative, A Rare Co-Existence.

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Abstract

Hidradenitis suppurativa (HS) is an autoinflammatory condition that attacks folliculopilosebaceous unit (FPSU), causing painful recurring inflamed nodules and abscesses mainly in intertrigenous areas of the body. Longstanding untreated HS increases the risk of anemia, cellulitis, scars, sepsis, fistula, damage of the lymphatic system and skin cancer(rare). Rare association was reported of HS with FSGS. Another rare literatures reported co exitance of HS with vasculitis. In our case we report rare coexistence of FSGS and vasculitis with HS patient.

Methods: We described our patient with HS in co-existence of Vasculitis, FSGS, new diagnosis of D2M and possibility to have bowel Vasculitis. vasculitis diagnoses were confirmed by a rheumatologist and was biopsy proven.

INTRODUCTION

HS is not common and has been reported mainly with two groups of disorders: autoinflammatory disorders, such as pyogenic arthritis and a group with folliculopilosebaceous structural disorders and hyperkeratosis, such as follicular occlusion syndromes (like in our case). HS may be associated with metabolic syndromes and inflammatory bowel diseases, but very rare to be associated with FSGS and vasculitis in one case, up to our knowledge no reported case has HS associated with vasculitis and FSGS, till now.

METHODS

We described our patient with HS in co-existence of Vasculitis, FSGS, new diagnosis of D2M and possibility to have bowel Vasculitis. vasculitis diagnoses were confirmed by a rheumatologist and was biopsy proven.

The diagnosis of HS was confirmed by a dermatologist using modified Dessau diagnostic criteria, which require typical morphology and location of the lesions and at least two flares in the past 6 months. A literature review was conducted through a MEDLINE, EMBASE and PubMed search using keywords “hidradenitis suppurativa,” “acne inversa,” “vasculitis,” “FSGS . Informed consent was obtained from the patients for publication.

Clinical presentation

A 36 years old, non-smoker man, obese, with BMI: 35kg/m2, presented to our hospital with newly diagnosed diabetes mellitus (D2M), proteinuria, picture of Inflammatory bowel syndrome (mucoid bloody diarrhea) and vasculitic skin rashes in both legs of 4 days duration.

On presentation he was feverish, RBS was 450mg/dl, HbA1C was 14.9, urine and blood ketones were positive with metabolic acidosis (Diabetic ketoacidosis, DKA) with picture of colitis, proteinuria +2, protein /creatinine ratio was 950mg/dl, with normal renal function tests.

He is a known case of Hidradenitis suppurative (HS) with atypical follicular occlusion tetrad (HS, scars of acne

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Received: 08-Feb-2025, Manuscript No. TAJOKD-4514 **; Editor Assigned:** 10-Feb-2025 **; Reviewed:** 24-Feb-2025, QC No. TAJOKD-4514 **; Published:** 08-Mar-2025, **DOI:** 10.52338/tajokd.2025.4514

Citation: Hoda Mohamed Hammoda. Newly diagnosed FSGS and vasculitis in a patient, known case of Hidradenitis suppurative, a rare co-existence. The American Journal of Kidney Diseases. 2025 February; 9(1). doi: 10.52338/tajokd.2025.4514.

conglubata, Multiple skin sinuses on the lower back which indicate the possibility of pilonidal sinus presence), in last 10 years, He was previously treated with various topical and systemic antibiotics with poor efficacy.

O/E

He was conscious, alert, looks dehydrated and toxic with sever abdominal pain 9/10, GSC: 15/15. Hypotensive and feverish. Lax neck, no palpable lymph nodes Skin examination: hydradenitis suppurative in the chest, both axilla, and groins (as described in pictures), both legs showed multiple erythematous follicular purpuric macules and papules some of them transformed to pustules (described in the picture), wound culture from that pustules was reported as no growth. The face showed wide areas of depressed scars of healed lesions of acne conglubata with few ice pic scars on the both upper cheeks and multiple large cobble stone comedons. Multiple skin sinuses on the lower back which indicate the possibility of pilonidal sinus presence.

Patient was admitted to medical ward at Hatta Hospital, kept NPO, stated on steroids, skin rash was biopsied which later reported vasculitis,

After stabilization of the patient, Ct +c abdomen was done which reflects the possibility of chron's disease, after couple of days, the patient was sent for upper and lower gastroenterology endoscopy to rule out inflammatory bowel disease, biopsy reported negative in three parts (stomach, duodenum and terminal ileum), for that thinking of bowel ischemia or bowel vasculitis was highly suspected.

As regard newly diagnosed D2M, patient was assessed by endocrinologist who started inulin and later improved with SGLT2 inhibitors. With marked improvement of the patient condition.

On review of his proteinuria in newly diagnosed D2M, and skin vasculitis with possible bowel vasculitis, the patient underwent renal biopsy, which revealed FSGS, The clinicopathologic findings are most consistent with a secondary form of FSGS rather than a primary podocytopathy. Possible etiologies include obesity-related glomerulopathy and arterionephrosclerosis, among others. There is no evidence of immune complex-mediated or paraprotein-related disease present.

Figure 1. HS hurley stage 3 of the chest: few inflamed nodules and abscesses covered with crustation. Multiple skin tunnels with thick and rode like scar tissue.

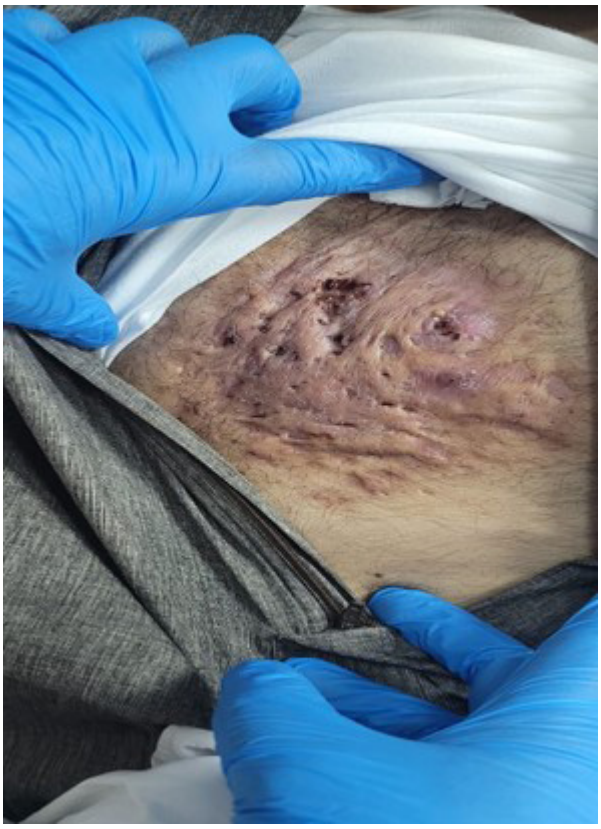


Figure 2. The right cheek showed wide areas of depressed scars of healed lesions of acne conglubata with few ice pic scars on the upper cheek and multiple large cobble.

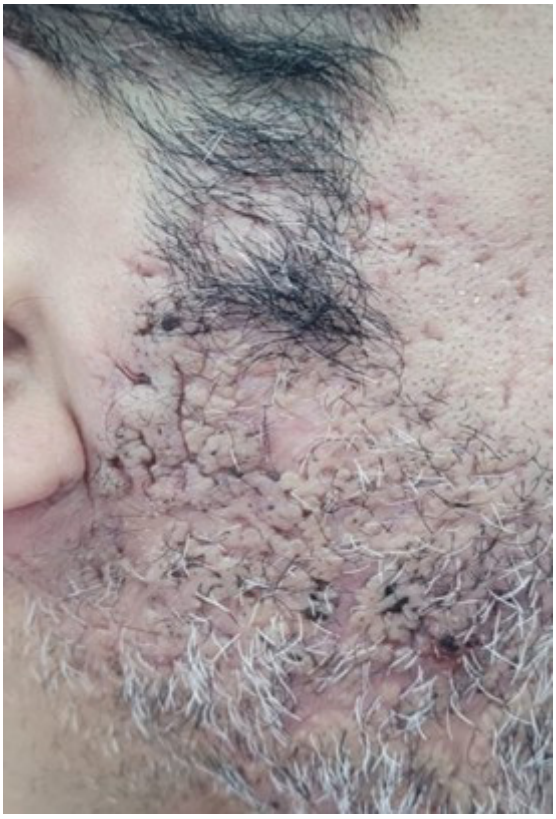


Figure 3. HS hurley stage 3 of both axillae: few inflamed nodules and abscesses. Thick interconnected rode like scar tissue studded with the openings of multiple skin tunnels.

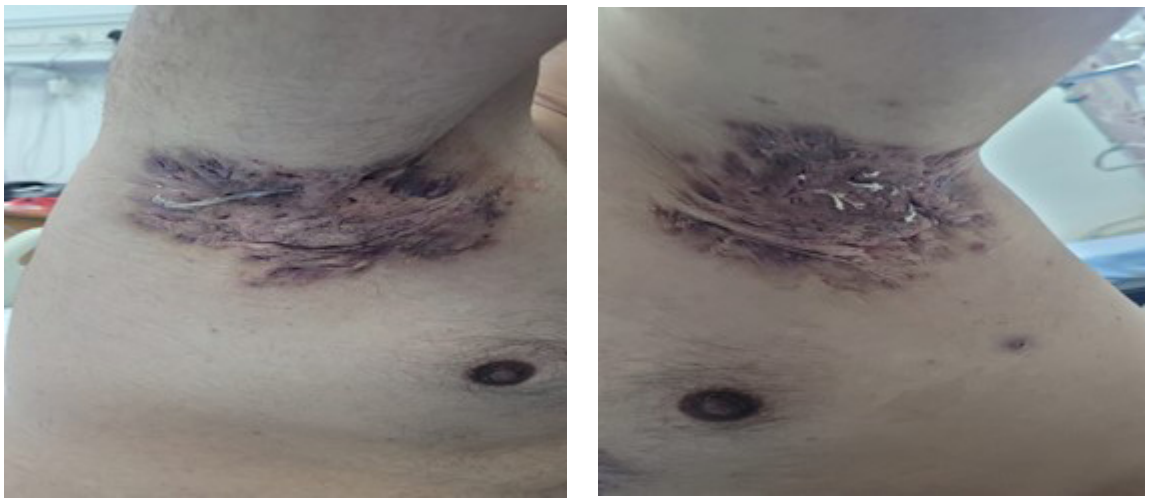


Figure 4. both legs showed multiple erythematous follicular purpuric macules and papules (palpable purpura).

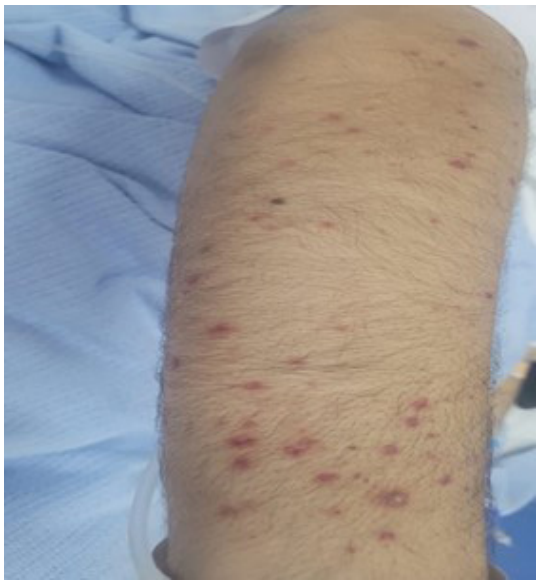
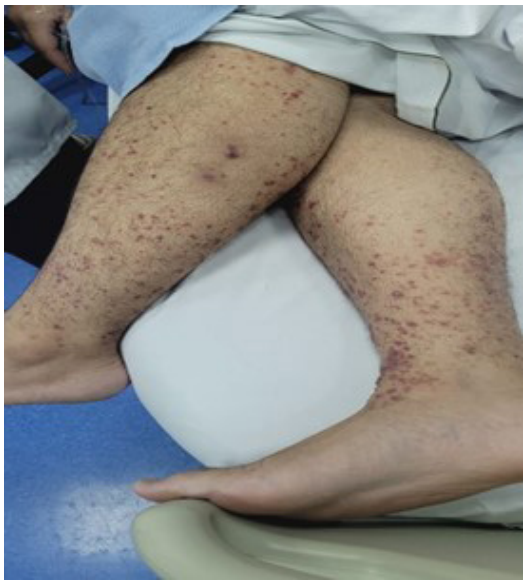



Figure 5. On the right lower limb: Multiple purpuric macules and papules with some of them transformed into pustules and coalesced together forming larger plaques.

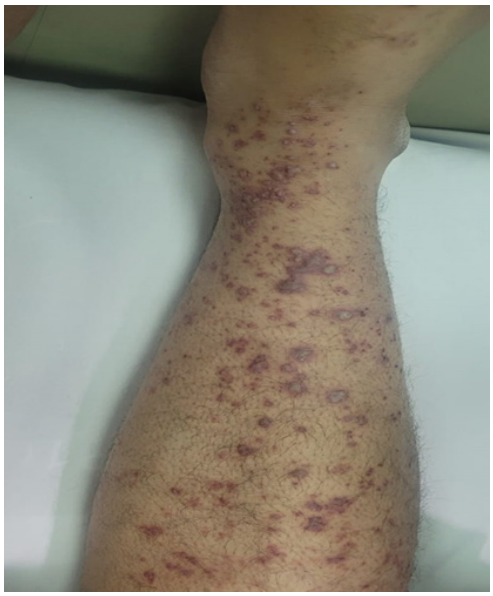
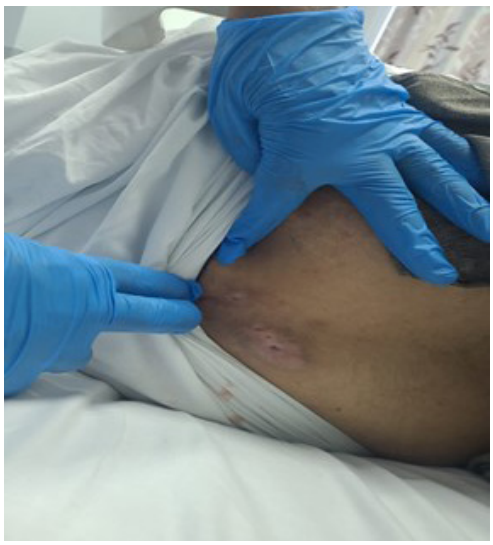


Figure 6. Multiple opening of the skin sinuses on the lower back, increasing suspicion of pilonidal sinus presence.



Laboratory findings

WBCS: 8.500/ul, HGB: 12.5g/dl, Plat.: 377, 000/ul. Urine routine: +2 protein, +4 Glucose, +4 ketones, RBCS: +1, Microalbumin/Creatinine Random Urine: 960mg/g

Urine Protein Creatinine Ratio: 650 mg/g, Protein Electrophoresis (Serum): No paraprotein band detected. Very Marked polyclonal increase in gamma globulins.

ESR: 40mm/1h , CRP: 123mg/l, ANTI Saccharomyces cerevisiae IgG abs. 78++ RU/ml < 20 , Saccharomyces cerevisiae IgA abs. >200+++ RU/ml < 20 (both are high), QuantiFERON -TB Gold Plus Blood: negative, RBS: 450mg/dl, HbA1c: 14.9, C-Peptide: 0.68 nmol/L, Islet cell antigen 2 AB (IA2): <2.5 U/mL, Islet cell AB (ICA): negative, Lipase: 27u/l, D-Dimer: 8.4ug/mL, Calprotectin: > 1000(high). Total protein: 10g/dl, Albumin: 2.9g/dl, Globulin: 7.2g/dl. CR: 0.8mg/dl, Urea: 24 mg/dl, K: 3.9 Mmol/L, Na: 128mmol/L, Glutamic decarboxylase AB (GAD65): negative, Anti Tissue Transglutaminase Blood: Negative, Thyroid Function Test (FT4, TSH): Normal, Hepatitis C Antibody Blood: Negative, HIV: Negative, Hepatitis B Surface Antigen Blood: negative. Antistreptolysin O: Negative, Rheumatoid Factor: negative, Anti-Nuclear Antibody (ANA) by IFA Blood: negative, ANTI-NEUTROPHIL CYTOPLASMIC Blood: Negative, CRYOGLOBULINS: Negative, ENA-Profile Blood, Venous: Negative, Anti Double Stranded DNA Blood, Venous: Negative, C3 and c4 were normal, Stool Routine; Stool blood: visible, stool mucus: Visible, stool pus: 5-10, Stool RBCs: 25-30. Stool Culture-Aerobic: NO SALMONELLA, SHIGELLA OR CAMPYLOBACTER ISOLATED. Helicobacter Pylori Antigen (Stool) Stool: Positive, Faecal Occult Blood (Faecal Immunochemical Test, FIT): Positive, Clostridioides Difficile Toxins A&B Stool: Negative, Gastrointestinal (GI) Panel – Film Array (Multiplex PCR): negative, Vasculitic skin rash, Wound Culture-: No growth.

Imaging and biopsy

CT Abdomen & Pelvis +C

- Wall thickening with mural enhancement seen involving long segment of the terminal ileum and extending to the ileocecal junction, corresponding mesenteric fat haziness and prominent mesenteric vessels noted, minimal free fluid noted mainly at the RIF, but no free intraperitoneal air, features are of terminal ileitis with Crohn disease on the top of the differential.

Histology Specimen GI Endoscopic Biopsy

A. Gastric biopsy: - No active inflammation seen - No intestinal metaplasia seen. - Giemsa stain: Negative for helicobacter pylori.

B. Duodenal biopsy: - No active inflammation seen - No significant pathology seen.

C. Terminal ileum biopsy: - No active inflammation seen - No significant pathology seen.

Skin Biopsy with Immunofluorescence (Vasculitis rashes)

A - Biopsy of skin - dermal vasculitis

B - Biopsy of skin for direct immunofluorescence - granular positivity for IgM, IgA and C3 in vasculitic dermal blood vessels Renal Biopsy Send Out Renal Biopsy Left;

- Focal Segmental Glomerulosclerosis, favor Secondary. - Global Glomerulosclerosis (5/40). - Interstitial Fibrosis, Moderate

Management

A Multidisciplinary Team, including Rheumatologist, Dermatologist, Nephrologist, Gastroenterologist, Endocrinologist and General surgery physicians were taking care of the patient, patient received steroids with Adalimumab and azathioprine as well, in addition to antiproteinuric agents (Ramipril and dapagliflozin).

Outcome and follow up

Patient was Advised for weight reduction strategies and also counseled as regard diet and exercise, continued on Dapagliflozin and Ramipril with Up titration of ramipril based on BP and proteinuria, Continued on Adalimumab and Azathioprine for Hidradenitis.

Patient was satisfied with that plane, and improving, as regard HbA1C reduced to 6, mucoid bloody diarrhea was completely improved and his vasculitis rash were disappeared, his proteinuria reduced from 950mg/g to 300mg/g, lastly HS still same with concern to switch to canakinumab.

DISCUSSION

Hidradenitis suppurativa (HS) is an autoinflammatory condition that attacks folliculopilosebaceous unit (FPSU), causing painful recurring inflamed nodules and abscesses mainly in intertrigenous areas of the body. There isn't a cure for HS, but treatment can help relieve symptoms. Longstanding untreated HS increases the risk of anemia, cellulitis, scars, sepsis, fistula, damage of the lymphatic system and skin cancer(rare). The reported prevalence of HS in older studies ranged widely from 0.4 to 4% (1).

Follicular occlusion disorders, inflammatory bowel diseases, spondylarthropathy and squamous cell carcinoma were the most common hidradenitis suppurativa comorbid diseases. Most of these diseases belong to the group of autoinflammatory disorders, where th17 cell cytokines seem to play a central role. (2)

the patients with HS may have increased incidence of certain diseases, such as metabolic syndrome and related comorbidities, as diabetes, obesity, insulin resistance, dyslipidemia, hyperglycemia and hypertension(3,9).

Rare association was reported of HS with FSGS. Another rare literatures reported co exitance of HS with vasculitis. In our

case we report rare coexistence of both FSGS and vasculitis with HS patient.

The prevalence of CKD in patients with HS was 6.3% compared to controls without HS 4.3%. The association of CKD was strongest in patients with HS who were the age of 60 years or older (16.9%), male (7.3%), obese (7.8%), have diabetes (12.5%), hyperlipidemic (13.3%), and had cardiovascular diseases (12.5%). (4)

In 2021, Akdoğan, et al, reported a rare association of HS and FSGS in a 24 years old male patient, with partially responsive to canakinumab (5). In the other hand, Demir et al, observed increase the risk of chronic renal failure in the patients have Hs in presence of amyloidosis with intervenient infections (6,10).

Vasculitis is uncommon disease which may affect small, medium and large vessels, its association with Hs is very rare. Aliturature published in 2019, who documented a series of five patients with HS associated with vasculitis, including one with Takayasu's arteritis (TAK), one with Behcet's disease (BD; variable vessel vasculitis subset) and three with granulomatosis with polyangiitis (GPA)) (7, 8).

Declaration of conflicting interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

Funding

The author(s) received no financial support for the research, authorship and/or publication of this article.

Informed consent

The patient has provided written consent for publication of the case report

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