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A Systematic Review of Follicular Psoriasis: A Unique but Under-Recognized Entity

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Abstract

Background: Follicular psoriasis (FP) is a rare and under-recognized subtype of psoriasis that affects hair follicles and can be frequently misdiagnosed due to its unique presentation. **Objective:** We aimed to analyze the frequently reported clinical, histological, and dermatoscopic features of FP, as well as their treatment options. **Methods:** We conducted a systematic review of the PubMed/MEDLINE database using the search terms "follicular" and "psoriasis." Fourteen studies were included yielding information on 44 patients (27 adults [61.4%] and 17 children [38.6%]). **Results:** Adult FP showed a female predominance (M:F = 1:2.7), frequent involvement of lower extremities (81.5%), association with metabolic syndrome including diabetes mellitus (22.2%), and a predilection for skin of color (SOC: White = 8:1). On the contrary, juvenile FP revealed male predominance (M:F = 1:0.6), frequent involvement of the trunk (41.2%), and exclusive involvement in skin of color (SOC:White = 11:0). In addition to its unique presentation, FP tends to be misdiagnosed due to its low incidence of concomitant psoriasis (31.8%), and rare personal (18.2%) or family history (6.8%) of psoriasis. Reported histopathological features include keratotic plugging, follicular parakeratosis with or without neutrophils, psoriasiform acanthosis, hypogranulosis, and neutrophilic infiltration of follicular epithelium. Dermatoscopic findings include folliculocentric lesions with normal appearing terminal hairs, perifollicular white scale, and various vascular structures. **Conclusion:** Once correctly diagnosed, patients with FP showed improvement or resolution of symptoms with various combinations of topical and systemic therapies. Understanding the demographic features and clinical presentations of FP can help address under-recognition of this clinical variant of psoriasis.

Keywords

follicular, psoriasis, skin of color, black skin, diabetes mellitus

Introduction

Follicular psoriasis (FP) is an inflammatory dermatosis in which histologic changes are localized/accentuated in the hair follicles. Follicular psoriasis was first described by Stankler et al in 1981 in a case series of 6 adult and 4 pediatric patients in Scotland.[1] The authors suggested 2 types; an adult form common in black, female patients with diabetes mellitus and a rarer juvenile form. Since this initial publication, occasional additional cases of FP have been reported.[2-11] The current literature emphasizes the distinction of this condition from the cutaneous manifestation of classical psoriasis variants and stresses diagnostic difficulty due to its apparent rarity and relatively unfamiliar clinical presentation. This study aims to analyze the clinical, histological, and dermatoscopic findings as well as treatment options of all FP cases reported in the literature.

Methods

Ethics Statement

This is a meta-analysis which does not require IRB approval. Not applicable.

A literature review using studies listed in the PubMed/MEDLINE database was performed in July 2022. The included keywords in the search strategy with Boolean terms were "follicular" AND "psoriasis". Cases were included if they described psoriasis with lesions in a follicular pattern (Figure 1). References within included articles were also reviewed to identify missed studies. The search strategy generated 194

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studies. After abstract review, 13 studies met the inclusion criteria. After review of references, an additional study was identified. Overall, 14 scientific articles detailing 44 patients with FP were included from 1981 to 2022 (Table 1). Extracted data points included age, sex, race/ethnicity/skin type, associated comorbidities, preexisting personal/family history of psoriasis, suspected triggering factors, clinical presentation, differential/previously rendered diagnoses, dermatoscopic and histopathologic findings and treatment to increase awareness of the entity and improve diagnosis of FP.

Results

Demographics

Of the 44 cases of FP reporting patient ages, 61.4% were adults (n = 27, range: 18-77 years) and 38.6% were pediatric (n = 17, age <18 years) (Table 1). Overall, the male to female ratio was 1:1.8 with a female predominance amongst adults (7:19) and a male predominance amongst children (5:3) (Table 2). Race/ethnicity was discernible for 20 cases (9 adults and 11 children). Where evaluable, FP showed a predilection for skin of color (SOC, n = 19) compared to lighter skin types (n = 1), yielding a SOC: White ratio of 19:1 (8:1 in adults and 11:0 in children). Of the adults with darker skin types, 5 were Black and 3 were Asian. Ten cases of FP were reported in Asian (Indian) children and 1 child of African descent.

Comorbidities

Of the 27 adult cases, diabetes mellitus (n = 6, 22.2%) was the most common comorbidity followed by hypertension (n = 2, 7.4%) and hyperlipidemia (n = 2, 7.4%). Out of 17 pediatric cases, two reported Crohn's Disease (11.8%).

History of Psoriasis and Association with Other Types of Psoriasis

While only 18.2% (n = 8) and 6.8% (n = 3) of cases reported having a personal or family history of psoriasis respectively, 31.8% of cases (n = 14) reported concomitant/pre-existing non-FP at the time of diagnosis of FP. Plaque and scalp psoriasis were the most associated subtypes. Among patients with non-FP, as a single or combined subtype, 6 had plaque psoriasis, 6 had scalp psoriasis, 2 had nail psoriasis, 2 had inverse psoriasis and a single patient had palmoplantar pustulosis (Table 1).

Triggering Events

Most cases of FP lacked a probable trigger. Three cases of suspected drug-induced FP were described. Two cases of FP triggered by infliximab for Crohn's disease[4] and 1 by pembrolizumab for non-small cell lung cancer[3] were reported.

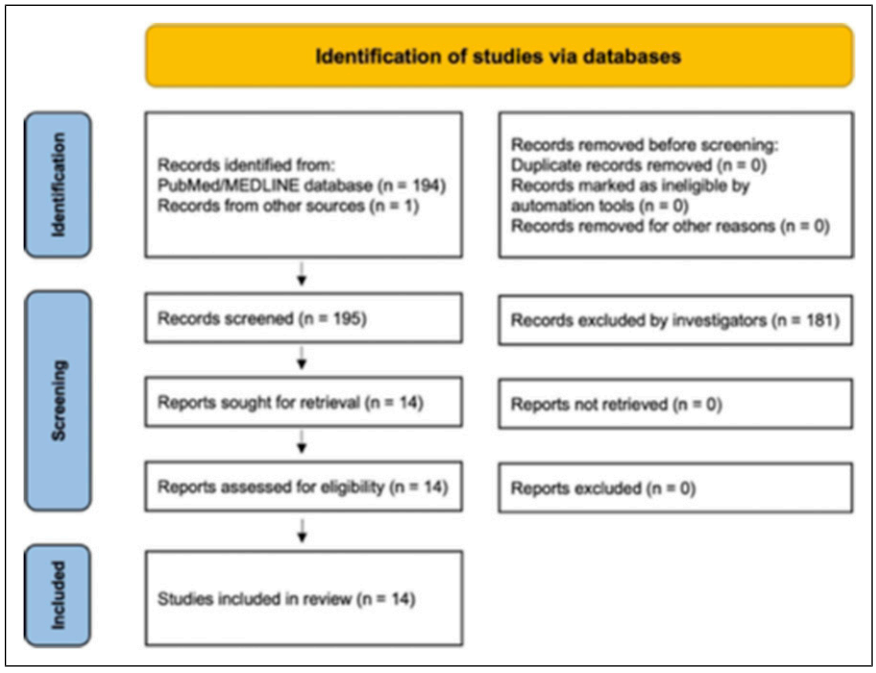

Figure 1. PRISMA flow chart. Results of a systematic review search strategy utilizing the PRISMA guidelines using studies listed in the PubMed/MEDLINE database. Keywords used to search included "follicular" AND "psoriasis". No additional filters were applied regarding language, dates, or type of publication.

Table 1. Characteristics of Included Cases of Follicular Psoriasis.

Author, year, country	n	Age	Sex (M:F)	Race/ ethnicity	Hx of pso	Fhx of pso	Comorbidities	A/w other types of psoriasis	Previous diagnosis	Treatment	Treatment response
Stankler,1981, Scotland[1]	10	<10 (4) 18-69 (6)	4:6	NR	NR	NR	NR	Plaque (1)	NR	Clobetasol, dithranol (2)	NR
Polysangam, 1997, USA[6]	5	23-73	1:4	Black (4), white (1)	NR	N	DM (3)	Scalp (2)	NR	Topical therapies	NR
Fatani, 2002, Saudi Arabia[12]	1	>18	NR	Middle eastern	NR	NR	NR	NR	NR	NR	NR
Thomas, 2010, United Kingdom[7]	1	18	1:0	Asian	Y	NR	NR	Scalp, plaque, nail	Lichen planopilaris	Narrow band UVB therapy	Resolution s/p 8 weeks
Arps, 2013, USA[13]	1	46	0:1	Black	N	NR	DM	N	Perforating disorder, sarcoidosis, verrucous LE, acute folliculitis	Modified Goeckerman, low dose MTX	Gradual improvement
Patil, 2014, India[8]	1	13	1:0	Indian	NR	N	NR	NR	NR	Emollients, MTX, vitamin D injection	Erythroderma improvement s/p 3 weeks, follicular lesions resolved s/p 8 weeks
Sathishkumar, 2015, India[14]	9	<18	NR	Indian	NR	NR	NR	NR	NR	NR	NR
Babino, 2016, Italy[9]	5	41-64	1:4	NR	Y (4)	NR	NR	Scalp (2), plaque (1), PPP (1)	NR	NR	NR
Nguyen, 2017, USA[10]	1	16	0:1	Black	N	N	NR	N	NR	Mometasone .1% ointment	Initial improvement
Behera, 2017, India[2]	1	34	0:1	Indian	NR	NR	NR	NR	Malassezia folliculitis, follicular lichen planus	Calcipotriol .005%/ clobetasol .05% ointment	NR
Souza, 2019, Brazil[11]	1	58	0:1	NR	NR	NR	DM	NR	PRP, KP + secondary infection	Acitretin	Improvement s/p 2 months
Scarfì, 2019, Italy[3]	1	69	1:0	NR	Y	NR	HTN	Plaque	NR	Prednisone, topical steroids	Resolution s/p 1 month
Magro, 2020, USA[5]	5	31-77	2:3	NR	Y (2)	Y (1), N (1)	DM (1), HTN (1), HL (2)	Plaque (2), scalp/ ear/nail (1)	Vasculitis, erythrodermic drug reaction, ACD, staph infection, folliculitis	Inhibitor, triamcinolone	Resolution
Goldberger, 2022, Israel[4]	2	13 (2)	1:1	NR	N (2)	Y (2)	Crohn's disease (2)	Inverse (2)	MSSA + folliculitis (2)	Discontinue TNF-α infliximab (2)	Improvement (1). Clearance with 1 episode of recurrence at 7 weeks, followed by gradual improvement 8 weeks later (1).

Abbreviation: n, Number of patients*; Y, Yes; N, No; M, Male; F, Female; NR, Not reported; Hx, history; Fhx, family history; pso, psoriasis; A/W, association with; DM, diabetes mellitus; HTN, hypertension; HL, hyperlipidemia; LE, lupus erythematosus; PRP, pityriasis rubra pilaris; KP, keratosis pilaris; ACD, allergic contact dermatitis; MSSA, methicillin-susceptible *Staphylococcus aureus*; MTX, methotrexate, TNF, tumor necrosis factor.

*Number (in parentheses) next to data point indicates number of patients within that report to which that data point applies.

Table 2. Comparison Between Adult Form vs Juvenile Form of Follicular Psoriasis.

Age group	Adult	Juvenile
Gender (M:F)	1:2.7	1:0.6
Skin type (skin of color:white)	8:1	11:0
HX of Psoriasis (%)	29.6	0
FHX of Psoriasis (%)	3.7	11.8
Comorbidities	DM (6), HTN (2), HL (2)	Crohn's disease (2)
Association with other types of Psoriasis	Plaque type (6), scalp (6), nail (2), palmopustular (1)	Inverse (2)
Location of lesions	LE (81.5%) > trunk (44.4%) > UE (33.3%) > axillae (22.2%)	Trunk (41.2%) > axillae (23.5%) > UE (11.8%) > LE (11.8%)
Treatment	Oral steroids, topical steroids, TNF-α inhibitors, acitretin, modified Goeckerman therapy, MTX	Emollients, MTX, topical steroids, discontinue infliximab

Abbreviation: M, Male; F, Female; Hx, history; Fhx, family history; DM, diabetes mellitus; HTN, hypertension; HL, hyperlipidemia; LE, lower extremities; UE, upper extremities; TNF, tumor necrosis factor; MTX, methotrexate.

Clinical Presentation

In adults, FP had been present between 1 month to 23 years at the time of reporting, while pediatric cases were present for 6 months to 4 years. Overall, most patients had involvement in multiple anatomic locations. The most common single location was the lower extremities (54.5%). The next most common site was the trunk (43.2%), followed by the upper extremities (25.0%), and the axillae (22.7%). Interestingly, the distribution of the lesions varied between the adult and juvenile forms (Table 2). In adults, FP most commonly involved the lower extremities (81.5%), followed by the trunk (44.4%), upper extremities (33.3%), and axillae (22.2%) (Figures 2A and B). By comparison, in pediatric patients, FP favored the trunk (41.2%), followed by the axillae (23.5%), upper extremities (11.8%), and lower extremities (11.8%) (Figures 2C and D). Associated joint involvement was reported in a patient with pembrolizumab which resolved after a prednisone taper despite continuing the medication.[3]

Differential Diagnosis

Ten cases included the initial differential diagnoses in their reports. The most common misdiagnosis was a folliculitis variant (n = 5). Other differentials included lichen planopilaris, allergic contact dermatitis, pityriasis rubra pilaris, keratosis pilaris, perforating disorder, vasculitis, erythrodermic drug reaction, sarcoidosis, cutaneous lupus, and superficial staphylococcal infection.

Dermatoscopic Findings

Three reports included dermatoscopic findings or photographs of active lesions. Folliculocentric lesions with normal appearing central terminal hairs, white-brown homogenous areas, and perifollicular white scaling were observed (Figure 3).[2] Additionally, multiple vascular structures including dotted, twisted, and/or glomerular/bushy capillaries were noted (Figure 3). Similar findings including central, normally appearing terminal hairs, perifollicular white scaling, and diffuse vascular structures were observed in 2 additional cases.[3,4]

Histologic Findings

Twenty seven cases reported skin biopsy findings. Follicular parakeratosis (88.8%) was the most commonly reported finding, while neutrophilic infiltration within the parakeratosis was noted in 40.7% (Figure 4). Other common findings in hair follicles included keratotic plugging (74.1%), follicular hypogranulosis 74.1%), follicular psoriasiform acanthosis (70.4%), follicular dilation (29.6%), and neutrophilic infiltration of the follicular epithelium (29.6%). No case reported follicular spongiosis. Around hair follicles, a perifollicular or perivascular lymphohistiocytic inflammation with neutrophils and rarely mast cells (88.8%) and perifollicular telangiectasia (18.5%) were identified. Interestingly, 2 cases reported absence of sebaceous glands.[5] In 81.5% of cases, interfollicular skin revealed typical changes of psoriasis.

Treatment

A variety of therapeutic approaches were tried for FP. The most prescribed therapeutic was topical monotherapy (n = 9). The topical agents employed included corticosteroids, dithranol, Vitamin D analogues, and emollients. Systemic medications prescribed included TNF-α inhibitors, oral antibiotics, oral prednisone tapers, methotrexate and acitretin. All cases in which a response to treatment was reported experienced significant improvement/resolution of FP. Drug induced cases resolved completely within 1-8 months. Although the infliximab-induced cases were treated by discontinuing the agent,[4] the pembrolizumab-triggered patient resolved after prednisone taper despite remaining on the medication.[3]

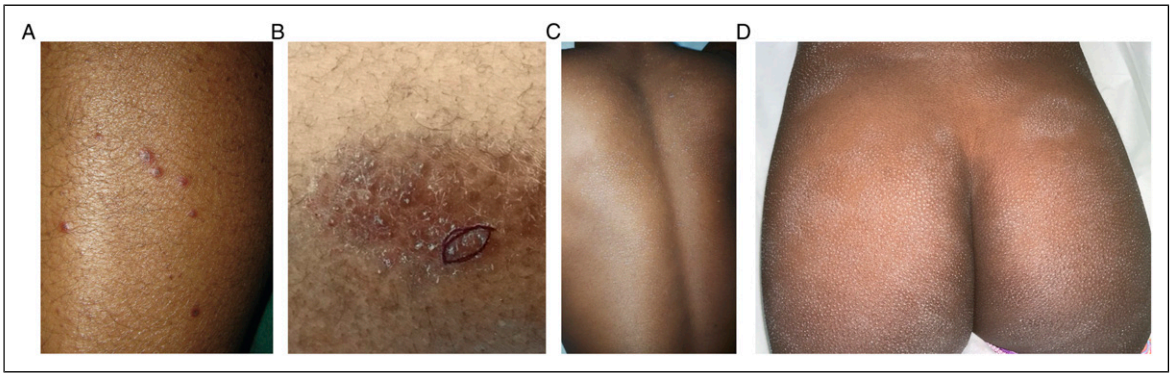

Figure 2. Clinical presentation of follicular psoriasis. (A) Adult form with multiple, discrete, erythematous scaly papules over the left lower leg (modified from *Indian J Dermatol Venereol Leprol* 2017; 83: 702-4).[2] (B) Adult form with an erythematous plaque with follicular accentuation and follicular scales on the thigh. (C) and (D) Juvenile form with follicularly based hyperkeratotic papules coalescing into discrete plaques.

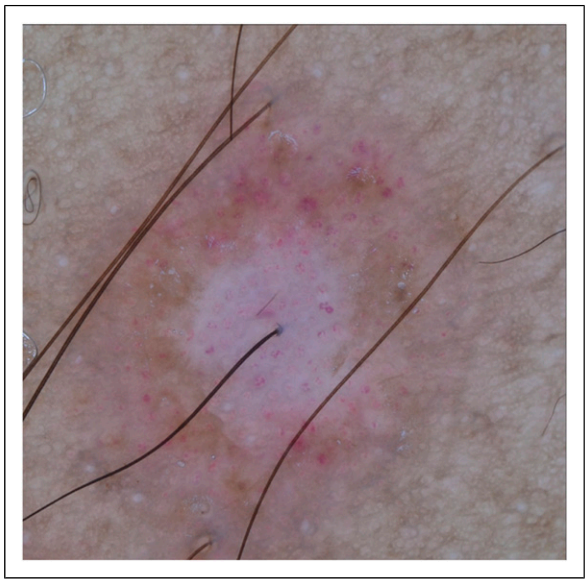

Figure 3. Dermatoscopic findings of follicular psoriasis: normal appearing central terminal hairs, white-brown homogenous areas, perifollicular white scaling, multiple red dots/dotted vessels, red globules, twisted red loops, and glomerular vessels/bushy capillaries (modified from *Indian J Dermatol Venereol Leprol* 2017; 83: 702-4).[2]

Discussion

Our review found that (1) FP was more commonly reported in adults (61.4%), although over a third of cases occurred in a pediatric age group (38.6%), (2) FP was more common in females in adults (73.1%), however, pediatric cases showed male predominance (62.5%), (3) 95% of those in whom race/ethnicity was reported were SOC patients but this demographic information was available in less than half of the included reports. Furthermore, the images provided for some of these (primarily adult) patients suggest lighter skin types. We are thus unable to confirm the previously suggested propensity of FP for darker skin types. It bears noting however, that race/ethnicity was reported for 11 of 17 pediatric cases (64.7%), and all of these were SOC patients. Ten of these were Asian (Indian) and 1 was Black. In pediatric cases of FP, there may be a predilection for patients with SOC.

Clinically, all patients presented with folliculocentric papules (Figure 2). Multiple areas of involvement were common in both adult and pediatric groups. In adults however, >80% had some involvement of the lower limbs (Figures 2A and B). Contrastingly, the lower limbs were only involved in 11.8% of childhood cases. Rather, a more central distribution was noted, with involvement of the trunk and/or axillae in 64.7% of cases (Figures 2C and D). Arthritis was only seen in the case of pembrolizumab-induced FP. Ten adult patients (37.0%) reported metabolic syndrome related disorders and diabetes was the most common of these (n = 6). Crohn's disease was reported in 2 pediatric patients.[4] While the number of cases is small, this correlates with the already described association of classical psoriasis with metabolic syndrome component diseases, reported between 20%–50%.[15]

FP tended to be misdiagnosed initially due to its unique presentations, a relatively low incidence of concomitant psoriasis, and rare personal or family history of psoriasis. In 11 cases, alternative diagnoses were initially rendered and folliculitis or classical folliculocentric diseases including lichen planopilaris, keratosis pilaris and pityriasis rubra pilaris were commonly suspected. Histologic examination and/or dermatoscopic examination appears helpful for increasing diagnostic accuracy of FP. The histological findings appear to confirm those of previous reports that lesions exhibit features typical of psoriasis vulgaris but accentuated in/limited to hair follicles. Findings include but are not limited to, keratotic plugging, follicular parakeratosis with neutrophils, psoriasiform acanthosis, and hypogranulosis.

Similarly, dermatoscopic findings are similar to plaque psoriasis but are seen in a folliculocentric pattern. Findings include unremarkable terminal hairs, perifollicular white scale, homogenous areas, and vascular changes (dotted/twisted/glomerular/bushy

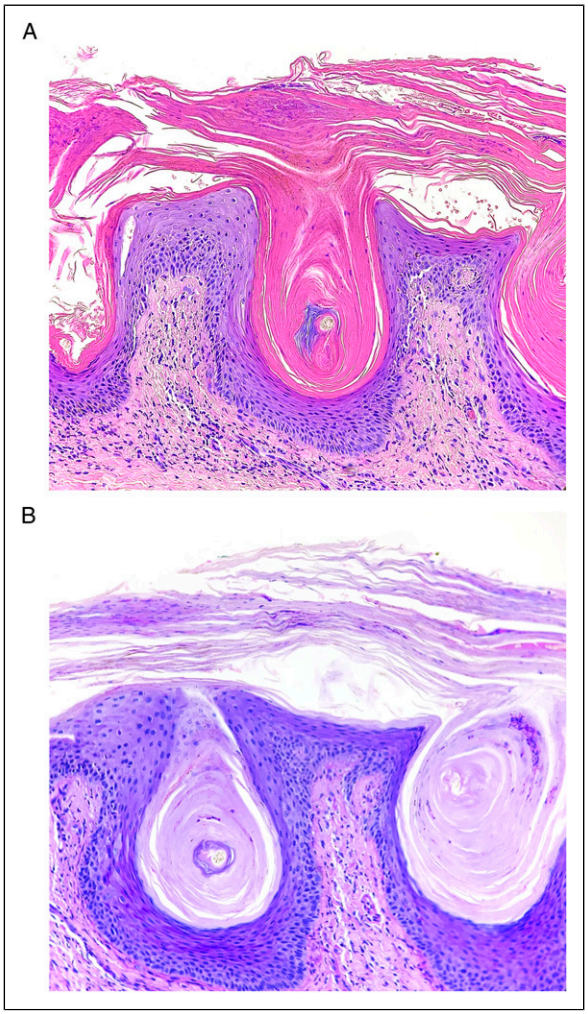

Figure 4. Histological findings of follicular psoriasis. (A) follicular plugging with parakeratosis containing neutrophils, mild follicular acanthosis, perifollicular telangiectasia, and a perivascular lymphohistiocytic infiltrate (H&E ×100 magnification). (B) PAS stain highlighting neutrophils within the follicular plugging and parakeratosis (H&E ×200 magnification).

vessels). These features have direct histopathologic correlates. Perifollicular white scale is histologically associated with perifollicular parakeratosis, perifollicular white homogenous areas with follicular/perifollicular hyperkeratosis and acanthosis, and vascular patterns with perifollicular telangiectasias.

Of the 16 cases that disclosed the regimen employed for treatment of FP, half reported successful management of the disease with topical monotherapy (n = 8). The remainder were treated with various systemic agents, light-based therapies, and combinations of various topical and systemic therapies to similar degrees of success. Of the 9 cases reporting a response to treatment, all observed notable improvement of symptoms if not complete resolution of FP. None of the cases reported recurrence of lesions. These findings suggest a relatively favorable prognosis for patients with FP with a relatively broad choice of therapeutic options.

Conclusion

This investigation marks the first review of the 44 total reported cases of FP from 1981 to 2022. The purpose served by this study was to clarify the demographic features of those most afflicted by FP, as well as further distinguish the rarer, but firmly present, pediatric manifestation of this disease. Our analysis reaffirms the importance of accurate diagnosis in cases of FP, with misclassification potentially leading to persistence of symptoms, in some cases, for decades. We hope that better understanding of this disease, its various clinical presentations, and the patient populations it most commonly impacts, will enhance rapid and accurate diagnosis and allow for appropriate and effective intervention. Limitations include missing demographic data and the retrospective nature of the study.

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.

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Ethical Approval

As a this review does not involve primary data collection or human subjects, formal ethics approval was not required. However, relevant ethical guidelines were upheld throughout the research process. By maintaining transparency, accuracy, and respect for the contributions of others, this work aims to contribute positively to the academic community and broader discourse within the field.

CME Credit

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Informed Consent

While this review does not involve direct interaction with human subjects, we recognize the importance of treating the works of authors, and the patients which they present, with the utmost respect and integrity. Every effort has been made to accurately attribute the ideas, findings, and intellectual contributions of authors and patients alike.

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