IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 23, Issue 11 Ser. 1 (November. 2024), PP 33-37
www.iosrjournals.org

An Incidental Finding Of A Tumour In A Case Of Pityriasis Rubra Pilaris- A Case Report

Dr Kantipudi Sree Swathi[1], Dr Shraddha Prabha[2], Dr Geeta Kiran A[3].
Junior Resident, Department Of Dermatology, Vydehi Institute Of Medical Sciences And Research Centre
Junior Resident, Department Of Dermatology, Vydehi Institute Of Medical Sciences And Research Centre
Proffessor And Head Of The Department, Department Of Dermatology, Vydehi Institute Of Medical Sciences And Research Centre

Abstract
We report a 49-year-old man who had a progressive rash that covered 90% of his total surface area in a period of 5 years. Confluent scaly erythematous plaques, which started on the upper back and spread to involve the scalp, face, chest, and limbs. Notable observations were subungual hyperkeratosis and onychogryphosis in all nails, along with follicular keratotic papules with waxy keratoderma on the palms and soles. Biopsy from skin lesions were suggestive of PRP.
A focal hypoechoic thickening at the base of the bladder was found during abdominal ultrasonography, which could indicate edema or a tumor. Histopathological examination of the removed tissue following TURBT revealed cystic urothelial nests that were consistent with Cystitis Cystica Glandularis (CCG), ruling out cancer. This case underscores the importance of a thorough investigation in patients with extensive dermatoses, highlighting a rare association between PRP and cystic bladder lesions.

Keywords: *Pityriasis Rubra Pilaris (PRP), Transurethral resection of the bladder tumor (TURBT)**,** Cystitis Cystica Glandularis (CCG)*

Date of Submission: 24-10-2024 Date of Acceptance: 04-11-2024

I. Introduction

An idiopathic papulosquamous disorder, pityriasis rubra pilaris (PRP) is characterized by follicular-based papules that coalesce into large confluent plaques, often progressing to erythroderma with distinctive areas of spared skin. While PRP is typically idiopathic, rare cases of paraneoplastic PRP have been reported, predominantly in the setting of solid organ malignancy.[1]

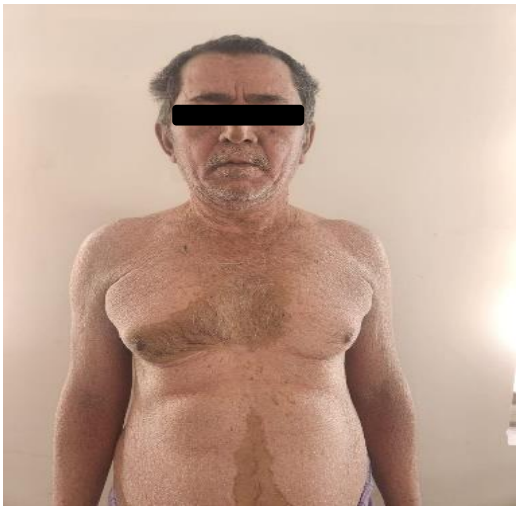
It has been noted that the Koebner phenomenon, UV exposure, and infections occur before PRP manifests. Reports of PRP's correlation with inflammatory arthritis, myositis, celiac sprue, and myasthenia gravis suggest that an autoimmune cause may be underlying. Internal cancers include renal cell, bronchogenic, and hepatocellular carcinomas have also been linked to HIV infection.[2]

We report a biopsy proven case of Erythrodermic PRP with suspected urinary bladder malignancy which was further evaluated.

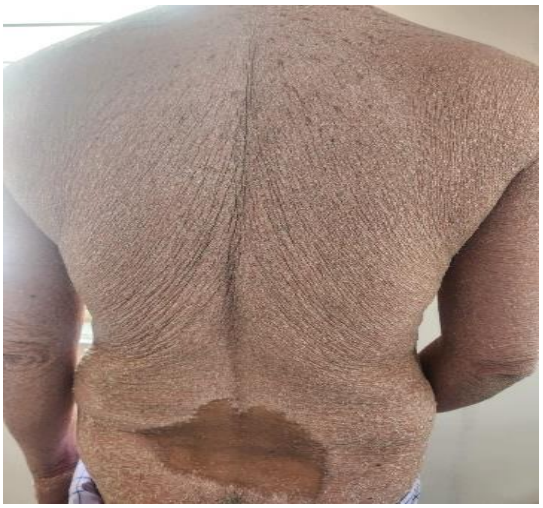
II. Case Report

A 49 year old male patient presented with a 5 year history of worsening of rash that began on the upper back and spread caudally to involve scalp, face, back, chest and extremities. Examination revealed confluent erythematous and scaly plaques involving approximately 90% of his total body surface area with islands of sparing over abdomen, lower back and thigh region. Follicular keratotic papules present over neck, chest, back and bilateral upper and lower limbs. There was waxy keratoderma of the palms and soles and nails showed subungual hyperkeratosis and onychogryphosis in all 20 nails of hands and feet. Routine investigations were normal and punch biopsy demonstrated ortho hyperkeratosis alternating with parakeratosis, follicular plugging and mild irregular psoriasiform epidermal acanthosis. Dermis shows mild superficial perivascular and perifollicular lymphocytic infiltrate and few dilated vessels seen, compatible with PRP. He was started on acitretin 25 mg daily. On routine USG abdomen and pelvis it was noted that there was a focal hypoechoic wall thickening at bladder base on the right side adjacent to the ureterovesical junction with a calcific focus as described and was suggestive of query oedematous UVJ/neoplastic etiology for which urology opinion was taken and was suggested TURBT (transurethral resection of bladder tumour) and patient was operated for the same. Its histopathological examination showed urothelial nests which are cystic and approximately of similar size. Occasional large sized

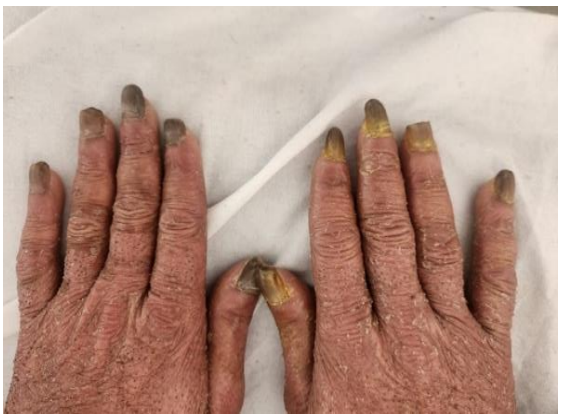
nests noted. Background shows oedema and dense lymphocytic aggregates. Urothelium is bland, features consistent of Cystitis cystic glandularis (CCG) and was negative for malignancy.



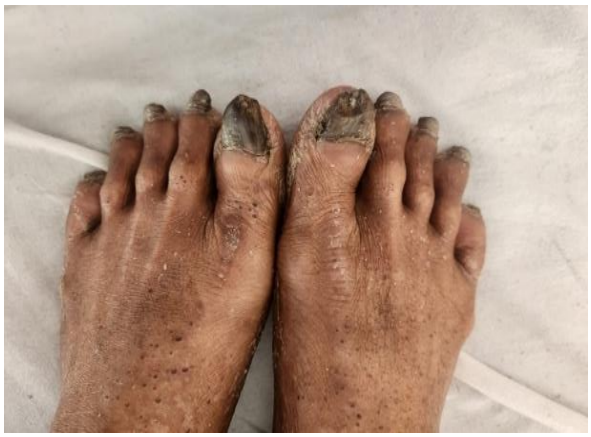
Diffuse erythematous plaques with scaling present over trunk and upper limbs sparing intermammary, right chest and midline of abdomen.



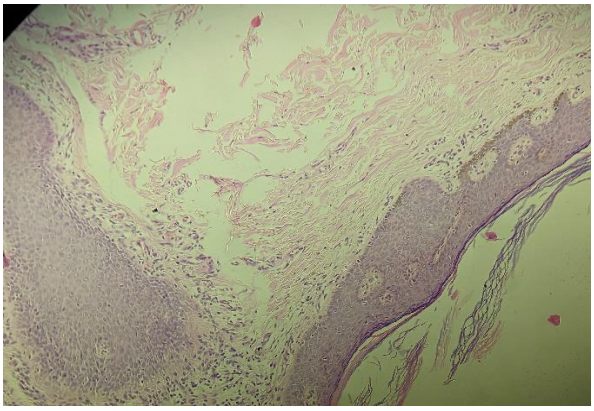
Diffuse erythematous plaques with scaling present over back sparing lumbosacral area.



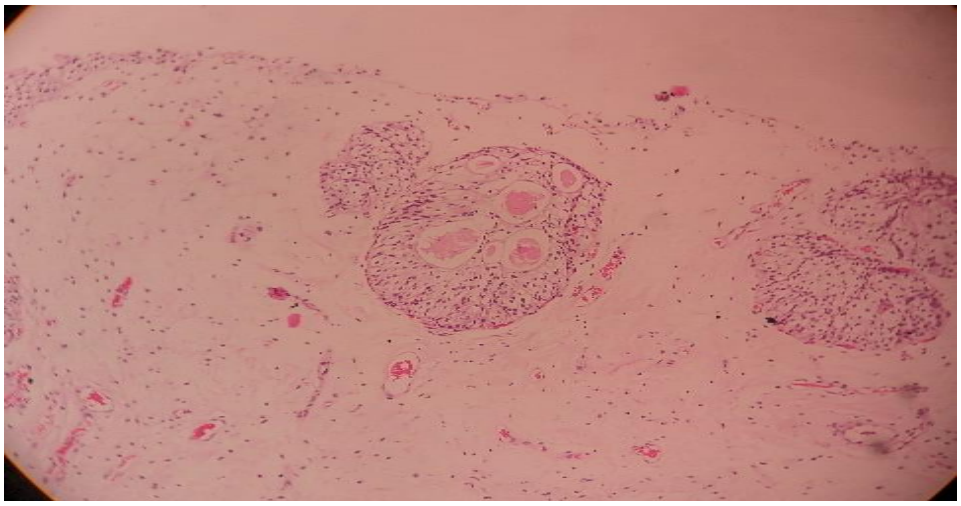
Yellow-brown discoloratation with subungual hyperkeratosis, thickening of nail plate present over all 10 fingernails and keratotic follicular papules present over B/L dorsum of hands.



Blackish discoloratation with subungual hyperkeratosis, thickening of nail plate seen over all 10 toenails with onychodystrophy of right great toe with keratotic follicular papules present over B/L dorsum of feet.



HPE of skin lesions on H and E staining showing ortho hyperkeratosis alternating with parakeratosis, follicular plugging and mild irregular psoriasiform epidermal acanthosis. Dermis shows mild superficial perivascular and perifollicular lymphocytic infiltrate and few dilated vessels seen, compatible with PRP.



HPE of excised tumor on H and E staining showing urothelial nests which are cystic and approximately of similar size. Occasional large sized nests noted. Background shows oedema and dense lymphocytic aggregates. Urothelium is bland, features consistent of Cystitis cystic glandularis and was negative for malignancy.

III. Discussion

Erythroderma is a generalised skin erythema that affects more than 90% of the body's surface area. Pityriasis rubra pilaris accounts for 1% of erythroderma patients with the incidence of one in 5000.[3] Basic characteristics shared by all subtypes of PRP include palmoplantar keratoderma and follicular papules that merge into distinct reddish-orange plaques with a nonadherent scale named as “nutmeg grater” or “exaggerated gooseflesh” with a hallmark feature of "islands of sparing".[4] Typically, it begins at the scalp and disperses cephalocaudally. PRP can occasionally be extremely diffusing, leading to erythroderma.[5]

It might be challenging to distinguish erythroderma secondary to PRP from erythroderma caused by other conditions, particularly psoriasis. In contrast to psoriasis, which displays silvery white scales and uniformly dispersed red dots on a backdrop of salmon red erythema, PRP has whitish keratotic plugs, a yellowish red background, and linearly oriented and dotted peripheral arteries. The follicular plugs, as well as the hyperkeratosis and parakeratosis in the perifollicular regions observed on histology, are correlated with the keratotic plugs and perifollicular scaling observed on dermoscopy. Histopathological observation of cutaneous capillary dilatation correlates with the linear and dotted vessels seen on dermoscopy.[6]

The characteristic features of histopathology include focal or confluent hypergranulosis, alternating orthokeratosis and parakeratosis in both vertical and horizontal directions (checkerboard pattern), and follicular plugging with perifollicular parakeratosis creating a shoulder effect. Thin dermal papillae, thick suprapapillary plates, wide rete ridges, and scant superficial perivascular infiltration, primarily of lymphocytes.[7]

There is not much information about the pathophysiology of paraneoplastic PRP. It is believed that immune reactions brought on by the underlying disease which cross-react with the skin could be the source of paraneoplastic skin disorders. In both familial and sporadic PRP, mutations in CARD14, a protein that controls keratinocyte responses to cytokine signals, have been found. A possible hypothesis is that when a CARD14 variation is present, immunological activity from a tumor may intensify the keratinocyte response, leading to a recurrence of a skin ailment that would normally be in remission.[1]

Regana et al. reported a case of PRP with previously undiagnosed malignancy with metastasis in the liver and primary tumor was not detected thus suggesting PRP as the initial manifestation of internal neoplasia.[8] L E Schwengle et al in his case report mentioned multiple eruptive seborrhiec keratosis associated with erythrodermic PRP.[9] A case of type II PRP associated with an ovarian cancer was reported in the 168 cases by Piamphongsant and Akarphant.[10]

Betinac T et al reported a case of rapidly progressing PRP as a possible initial cutaneous symptomof a previously undiagnosed laryngeal carcinoma.[11]

Cystitis glandularis is a proliferative disorder of the urinary bladder, which tends to be associated with glandular metaplasia of the transitional cells lining the bladder. In addition to being benign simulators of invasive bladder cancer, cystitis cystica and cystitis glandularis frequently coexist with associated lesions.[12]

An ultrasound scan of the renal tract can demonstrate a polypoidal thickening of the wall of the urinary bladder, usually in the trigone area. In more extensive cases, this thickening can be present throughout the entire bladder. If the ureteric orifices are obstructed, there may be evidence of hydroureter and hydronephrosis. A CT scan may show a hypervascular polypoid mass within the urinary bladder, while an MRI scan can demonstrate a hyperintense vascular core with a surrounding low-intensity signal.

Diagnosis of cystitis is typically made based on histopathology examination and immunohistochemistry staining studies of biopsy specimens or transurethral resection specimens of the urinary bladder lesions. Microscopy pathology examination of the specimens generally demonstrates:

(a) Abundant urothelial von Brunn nests, which often exhibit a vaguely lobular distribution of invaginations, evidence of non-infiltrative growth, and variable connection to the surface.

(b) Gland-like lumina with columnar or cuboidal cells in cases of cystitis glandularis.

(c) Cystically dilated lumina or cystic cavities filled with eosinophilic fluid in cases of cystitis cystica.

(d) Most cases of cystitis demonstrate coexistence of both patterns.

(e) Cells lack significant atypia, mitotic activity, stromal reaction, and muscular invasion, although degenerative atypia is occasionally present.

They tend to exhibit positive immunohistochemistry staining for various markers, including GATA3, CK7 (full thickness), CK20 (umbrella cells), p63 (basal cell layer), uroplakin II/III, thrombomodulin, beta-catenin (membranous), and E-cadherin. On rare occasions, cystitis cystica and cystitis glandularis can coexist with urothelial carcinoma. In 75% of instances, bladder dystrophy and pelvic lipomatosis are linked to CCG. These illnesses operate as risk factors for the development of disease and increase the possibility of malignant transformation into adenocarcinoma.[12] Therefore, pathologists must thoroughly examine bladder lesion specimens to ensure there is no synchronous malignancy.

The treatment of cystitis cystica involves removing the source of irritation or bladder inflammation, such as foreign bodies, long-term urinary catheters, or vesical calculi, as well as performing transurethral resection of the bladder lesion or lesions.[13]

Often recalcitrant to treatment, PRP has no US Food and Drug Administration (FDA)-approved treatments. Topical corticosteroids, topical calcineurin inhibitors, keratolytic medicines, and emollients are common treatments for mild PRP. Systemic therapy, including oral retinoids and methotrexate, has shown promise for managing symptoms and reducing inflammation in more severe cases. Biologics commonly used in the management of psoriasis, such as tumor necrosis factor alpha (TNF alpha) inhibitors, secukinumab, and ustekinumab, have recently been used for the treatment of PRP. Compared to traditional PRP, paraneoplastic PRP (pPRP) is less responsive to typical therapies and is particularly challenging to treat without addressing the underlying malignancy.[2]

IV. Conclusion

Paraneoplastic PRP can signal underlying malignancies and is challenging to treat, often requiring management of the associated cancer. Cystitis glandularis involves bladder lesions that can mimic cancer requiring careful histopathological evaluation. Treatment for PRP and cystitis glandularis varies with PRP often requiring systemic therapy and cystitis treatment focusing on removing irritants and resection.

References

[1] Lamberg, O., Cao, S., Sabater-Geib, S., Lowe, L., & Elder, J. (2021). A Case Of Recurrent And Paraneoplastic Pityriasis Rubra Pilaris. Jaad Case Reports, 12, 74-76.

[2] Mehta N, Coates Mm, Miles Ja, Miedema J, Blasiak Rc. A Case Of Paraneoplastic Pityriasis Rubra Pilaris. Jaad Case Rep. 2023 Jul 13;39:125-129. Doi: 10.1016/J.Jdcr.2023.06.045. Pmid: 37680569; Pmcid: Pmc10480444.

[3] Khan, I., & Khalil, U. (2005). Erythroderma Due To Pityriasis Rubra Pilaris. Journal Of Pakistan Association Of Dermatology, 31(2), 233-235.

[4] Greiling Tm, Brown F, Syed Ha. Pityriasis Rubra Pilaris. [Updated 2024 Apr 30]. In: Statpearls [Internet]. Treasure Island (Fl): Statpearls Publishing; 2024 Jan

[5] Selma E, D. Dd, Ahmet M, Özkay Ö, Nuran Sa, N. Da. An Acute Onset Erythrodermic Adult Pityriasis Rubra Pilaris Case And Response To Treatment With Methotrexate. J Turk Acad Dermatol.2014 Mar;8(1):0-0.

[6] Vinay K, Kumar S, Radotra B. Dermoscopy Of Erythrodermic Pityriasis Rubra Pilaris. Indian Dermatol Online J [Internet]. 2019;10(4):500.

[7] Kuvvarapu Ak, Gopal Kvt, Raju Pvk, Rani Br. Atypical Adult Onset Pityriasis Rubra Pilaris: A Rare Chronic Form Of Pityriasis Rubra Pilaris. Int J Res Dermatol [Internet]. 2022;9(1):55.

[8] M. Sänchez-Regaña, F. López-Gil, M. Salleras, P. Umbert, Pityriasis Rubra Pilaris As The Initial Manifestation Of Internal Neoplasia, *Clinical And Experimental Dermatology*, Volume 20, Issue 5, 1 September 1995, Pages 436–438.

[9] Schwengle Le, Rampen Fh. Eruptive Seborrheic Keratoses Associated With Erythrodermic Pityriasis Rubra Pilaris. Possible Role Of Retinoid Therapy. Acta Derm Venereol [Internet]. 1988 [Cited 2024 Jun 25];68(5)

[10] Piamphongsant T, Akaraphant R. Pityriasis Rubra Pilaris: A New Proposed Classification. Clin Exp Dermatol [Internet]. 1994 [Cited 2024 Jun 25];19(2):134–8.

[11] Batinac T, Kujundzić M, Peternel S, Cabrijan L, Troselj-Vukić B, Petranović D. Pityriasis Rubra Pilaris In Association With Laryngeal Carcinoma. Clin Exp Dermatol. 2009 Dec;34(8):E917-9.

[12] Fariña La. Re: Pelvic Lipomatosis Associated With Cystitis Glandularis And Adenocarcinoma Of The Bladder. J Urol 1992;147(5):1380.

[13] Kodzo-Grey Venyo A. Cystitis Cystica And Cystitis Glandularis Of The Urinary Bladder: A Review And Update. J Clin Res Rep [Internet]. 2022;11(1):01–22.