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A Case Report on Pityriasis Rubra Pilaris (PRP) and Multisystem Inflammatory Syndrome in Children (MIS-C)

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ABSTRACT

Pityriasis rubra pilaris (PRP) is a rare inflammatory papulosquamous skin disease of unknown etiology. There are six distinct types of PRP. A higher incidence was reported within the pediatric group, but treatment remains largely empiric, based on case reports and series. The condition is often refractory to treatment, especially initially, with topical corticosteroids and oral Acitretin the more commonly employed agents. biologic agents approved for psoriasis have been used with good effect in PRP. Multisystem Inflammatory Syndrome in Children (MIS-C) is a new disease entity that evolved as a result of the coronavirus disease 2019 (COVID-19) pandemic. Due to our incomplete understanding of the occurrence of MISC, new diagnostic and therapeutic issues have emerged. The American college of rheumatology (ACR) has recommended the use of intravenous immunoglobulin (IV IG) and/or high-dose corticosteroids as first-line treatment in these patients. In some cases of severe patients may receive such as a second dose of IV IG, Anakinra or Infliximab and Aspirin. Here we report the case of a 4-year-old boy presented with rashes. We describe the successful use of IV IG, corticosteroids, and Aspirin for treatment of MIS-C and PRP.

Keywords: Pityriasis rubra pilaris, MIS-C, IV IG

INTRODUCTION

Pityriasis rubra pilaris (PRP) is an uncommon type of inflammatory skin condition marked with the aid of using palmoplantar hyperkeratosis and erythematous salmon-coloured plaques with sparing islands.[1] Although the cause is unknown, there has been recent speculation that it may be caused by immune system dysregulation, where there is an aberrant response to antigenic stimuli.[2] PRP manifests with some similarities to psoriasis but has diverse histopathologic characteristics.[1,3] Histopathology can be non-specific, although in typical cases it reveals psoriasiform hyperplasia, a persistent granular layer, and alternating orthokeratosis and parakeratosis in vertical and horizontal planes.[4] It has peaks of age of onset withinside the first and 5th to 6th decades, affecting each ladies and men equally.[5,6] There are five different forms of PRP, according to Griffiths' classification.[7] Type I - classic adult type, Type II - atypical adult type, Type III - classic juvenile type, Type IV - limited juvenile type, Type V -

atypical juvenile type. More recently, Miralles et al. added an HIV-related type (Type VI) to this classification system.[8] In India and the United Kingdom, the incidence was estimated to be one in 500,000 to one in 5000 patients, respectively. [4,9,10] But the pediatric group was found to have a higher incidence.[11] Type 2 and Type 5 have longer disease duration than Type 1 and Type 3. Any type may exhibit follicular hyperkeratosis. About 5% of cases of juvenile PRP are Type 5 abnormal instances. It typically appears throughout the first ten years of life.[4] Topical corticosteroids, vitamin D supplements, oral retinoids, methotrexate, and cyclosporine are examples of conventional therapies. [4,12,13] Biologics have recently become more popular because of their manageable side effects.[4]

The unusual complication known as Multisystem Inflammatory Syndrome in children (MIS-C) is thought to be related to COVID-19 and is brought on by an overactive inflammatory response and unidentified immunological dysregulation. [14-16] According to several reports children may still develop MIS-C even if their case of SARS-CoV-2 is asymptomatic and mild.[17-18] The definition of MIS-C is based on the following principal elements: age, presence of fever, elevated levels of inflammatory markers, the involvement of more than two organ systems, timing of COVID-19 infection or exposure, and exclusion of other diagnoses.[19]

A multisystemic inflammatory reaction (MIS-C) indicated a temporal association between infection and onset of symptoms during the early months of the COVID-19 pandemic in certain children.[20] According to research that have been published, MIS-C may have a high fever and non-specific symptoms such as nausea, vomiting, headaches, and exhaustion 2 to 6 weeks after contracting the SARS-CoV-2 virus, as well as in some cases conjunctival hyperemia and a rash that resembles Kawasaki syndrome.[21] MIS-C in genetically predisposed infants is caused by a complicated process that includes the presentation of SARS-CoV-2-specific antigen to autoreactive T cells, superantigen like viral structures, cross-reactive SARS-CoV-2-specific antibodies, and imbalanced cytokine responses.[22] The primary and effective treatment for Kawasaki disease is immunoglobulin g (IV IG). while some MIS-C children may improve with care and treatment.[21] The first-tier treatment in MIS-C that is most usually administered is IV IG. However, IV IG therapy entails high fluid infusions (40 ml/kg), which could exacerbate myocardial dysfunction in the early stages of the disease.[23] Following IV IG therapy, several inflammatory markers are reduced in MIS-C patients. In fact, IV IG has been found to prevent T cells, monocytes, dendritic cells, and endothelial cells from becoming activated in MIS-C. Additionally, MIS-C showed non-specific activation of B cells with autoimmune markers, and IVIG could check B cell activation. Even though superantigen-mediated lymphocyte activation has been demonstrated to be suppressed by IV IG.[24] Because of the rare reporting of the disease in childhood, we are reporting this classical case of childhood onset PRP and MIS-C.

CASE PRESENTATION

A 4-year-old male child was presented with complaints of fever for 2 days followed by rashes over palm, soles and around lips, redness and swelling all over body and was admitted in department of paediatrics. Rashes were spread all over the body, initially papular lesion, later became scaly and peeling of skin associated with severe itching and consulted many dermatologists and was prescribed with moisturizing and steroidal ointment for rashes. There was no history of cold, loose stool, vomiting, ulcers over mouth, conjunctival congestion. He has a family history of paternal cousins having allergic history. No prior food or drug allergy was identified.

At the time of admission, the child was active and alert. He was febrile (100.5°F) and had scaly and erythematous lesions

(papules and plaques) over body with Keratosis at Palms, Soles, fissuring and oedema. There was no mucosal involvement, oral cavity was normal and no lymphadenopathy or organomegaly was present. Child was started on IV antibiotics (Ceftriaxone) with provisional diagnosis of Psoriatic erythroderma, staphylococcus-scalded skin syndrome (SSSS) after sending blood investigations. Child continued to have high grade fever spikes, along with flushing of face, erythema.

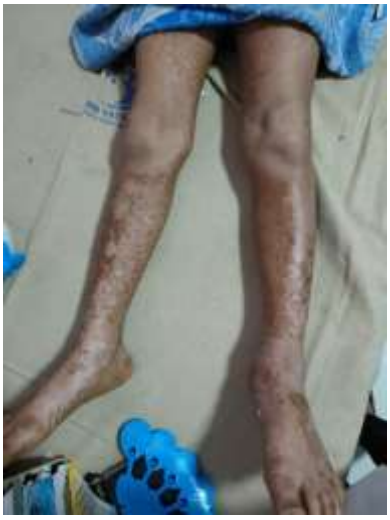

Fig. 1a Rashes over both lower limbs

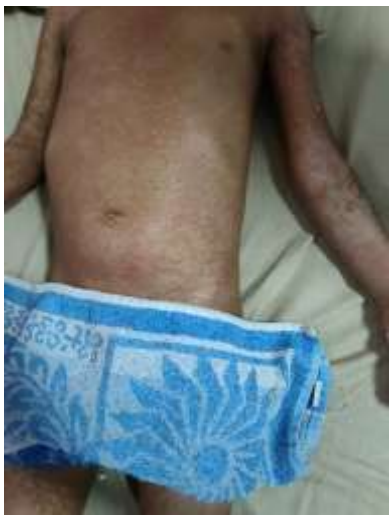

Fig. 1b Rashes over both upper limbs and body

FIGURE 1: PRP LESIONS OVER BOTH LOWER LIMBS (1a) AND OVER UPPER LIMBS AND BODY (1b)

His initial laboratory data showed declined Haemoglobin (10.1 gm/dL), Serum Globulin (2.3 gm/dL), and Total Protein (4.9 gm/dL). The elevated parameters include Total Count (28540 cells/cumm), CRP (245.2 mg/L), D-Dimer (15,859.60 ng/ml), BNP (406 pg/ml), serum LDH (367 U/L), Procalcitonin (4.72 ng/ml), Covid IgG (>200 BAU/ml). Peripheral Smear Study showed mild neutrophilic leukocytosis and thrombocytosis.

Skin biopsy of epidermis showed alternating orthokeratosis and parakeratosis in both vertical and horizontal directions. Focal hypergranulosis, thick suprapapillary plate, narrow dermal papillae and follicular plugging are noted. Superficial dermis showed mild to moderate perivascular lymphocytic infiltration and was diagnosed with Pityriasis rubra pilaris (PRP) on dermatology consultation.

On ID (Infectious Diseases) consultation, Since the child continued to have high grade fever with erythema, exfoliation, oedema, the child was diagnosed with MIS-C/atypical Kawasaki based on following criteria.

Criteria fitting MIS-C are as follows:

1) Age- 4 years
2) Fever > 3 days
3) Multi system involvement – Rash, elevated D-Dimer
4) Elevated CRP, Procalcitonin
5) Blood C/S-sterile, other infections ruled out
6) Covid IgG Positive

The patient was treated under the guidance of pediatricians and dermatologist of the hospital. The MIS-C was managed with Intravenous Immunoglobulins (IV IG). Here IV IG was administered at 12 ml/hr for 15 minutes, 24 ml/hr for next 15 minutes, 48 ml/hr for next one hour, 75 ml/hr for next one hour, totally of 5 days. The steroidal medications administered for the management of MIS-C and PRP included Tab. Omnacortil (Prednisolone) 20mg twice daily for three days and Inj. Methylprednisolone 40mg once daily for three days. Antithrombotic treatment included Tab. Ecosprin (Aspirin) 75mg ¾ once daily for three days. The topical treatment included Cetaphil with Momate (Mometasone) combination cream and

Liquin Cream (Liquid light paraffin and White soft paraffin) for local application twice daily. The child was treated with following antibiotics Inj. Monocef (Ceftriaxone) 750mg twice daily (2 dose) and Inj. Amikacin 150mg twice daily (1 dose), and the above antibiotics were stopped and then upgraded to Inj. Piptaz (piperacillin tazobactum) 2gm thrice daily for five days (16 doses), Inj. Linezolid 200mg given thrice daily for 5 days. Inj. Avil (Pheniramine) 7.5mg as stat and Syp. Allegra (Fexofenadine hydrochloride) 5ml twice daily for one week was also given. Syp. Calpol (Paracetamol) 6ml six times daily, Syp. Zincovit (multivitamin) 5ml given twice daily were also included in the treatment plan. The abnormal parameters were normal after treatment and vitals is stable with no new lesions and hence being discharged.

Advice on discharge included Syp. Zincovit (Multivitamin) 5ml twice daily for two weeks, Tab. Omnacortil (Prednisolone) 20mg twice daily for five days, Tab. Ecosprin (Aspirin) 75mg for ¾ once daily for six weeks, Tab. Junior Lanzol (Lansoprazole) 15mg once daily for 5 days, Syp. Allegra (Fexofenadine hydrochloride) 5ml for 5 days, Sorvate (Calcitriol) ointment for once daily on trunk papules, Momate cream (Mometasone) for local application once daily on trunk papules, Liquin cream (Liquid light paraffin and White soft paraffin) for local application twice daily over extremities, Cetrilak (Cetrimide) mild lotion for local application over scalp twice weekly, Cetaphil lotion for local application twice daily, Mupirocin cream for local application on fissure and biopsy site.

DISCUSSION

Pityriasis rubra pilaris is an unprecedented papulo-squamous ailment of idiopathic etiology. Because PRP is a rare disease, its aetiology and pathophysiology are still poorly known, and the condition's management mainly rely on prior case reports. There is a distinct lack of data covering paediatric patients. There are no double-blind, randomised, controlled studies that include treatment alternatives.[1] Therefore, there is no evidence for standard care. Evaluation of any therapy's effectiveness is so debatable. Given that patients with PRP frequently respond favorably to retinoid therapy, some writers think that aberrant vitamin-A metabolism is manifested in these patients. Others connect immunological dysregulation to the illness. According to a third view, juvenile PRP and guttate psoriasis are both superantigen-mediated diseases.[25] Therefore, the sources of recommendations were limited to retrospective case series and case reports, levels four and five of evidence. A few therapeutic strategies for treating PRP has been reported. In addition, some PRP instances recover on their own without the need for therapy.[3] More than 20 different medications have been used to treat the disease, according to research done in 1985 by Fox et al. [26] The second-line medication of choice historically has been methotrexate, but systemic retinoids are frequently utilized as the first-line therapy. The use of immunosuppressive drugs such azathioprine or cyclosporin A, stanozolol, phototherapy, extracorporeal photochemotherapy, fumaric acid esters, and calcipotriol are among the further treatments. Tumour Necrosis Factor (TNF)-α. is a pro-inflammatory cytokine that is bound by the neutralizing, chimeric monoclonal antibody infliximab.[27] One of the treatment techniques used in PRP is phototherapy. Given that our patient has a history of photo aggravation, it should be avoided in his case. High-potency corticosteroids, tar, calcipotriene (calcipotriol), keratolytics, and tretinoin are examples of topical medications that can be used in conjunction with systemic therapy.[28]

The regimens utilized for MIS-C patients are comparable to those for those with Kawasaki disease because of the syndrome's recent start and its similarity to that illness. MIS-C treatment guidelines were recently released by ACR.[29] IV IG and/or high-dose corticosteroids have been suggested by the

ACR as first-line treatments for these patients. Between 30 and 80 percent of patients do not react to IV IG as a monotherapy and may require further immunomodulatory therapy to reduce inflammation. [30,31] Less than 15% of patients with MIS-C from Kawasaki disease show resistance to IV IG therapy. In some situations, patients with severe conditions may get methylprednisolone (10–30 mg/kg/day for 3–7 days, followed by a gradual decrease of oral prednisolone) and aspirin. Other treatments may also be given, such as a second dose of IV IG, anakinra, or infliximab. [32,33] There are few clinical practice recommendations regarding treatment for MIS-C by multiple organizations. Positive outcomes in therapy regimens utilizing IV IG and corticosteroids were reported by Ramcharan et al. [34] Like this, IV IG, corticosteroids, and anticoagulants all produced positive results in our experience. Our report provides further evidence of this observation and suggestions for treatment of PRP and MIS-C. The child was started on injection IV IG, corticosteroids and anticoagulants following the child became afebrile and vitals stable. Repeated investigations showed decline in inflammatory markers, hence being discharged.

CONCLUSION

PRP is one of the rare dermatological disorders that may go undiagnosed for years. A thorough history and physical examination with clinicopathological association is therefore necessary. In addition, some individuals with PRP type 5 have familial variations, such as those who have a CARD14 gene mutation. Family counselling and genetic testing are therefore advised for PRP type 5 individuals. Due to the characteristic clinical symptoms, doctors must diagnose patients, most commonly dermatologists and paediatricians. The stress experienced by both the clinician and the patient would be reduced if both were aware of the disease's clinical characteristics. The improvement of PRP patients' quality of life may be improved by the development of a standardized treatment. When the prevalence of COVD-19 has peaked, every child who has a fever should have a thorough clinical evaluation for probable multiple organ-system involvement and a well-defined epidemiological history, with an emphasis on those who have significant abdominal discomfort and/or skin or mucocutaneous lesions. In order to improve patient care, COVID-19 related with MISC needs more research and investigation. A thorough follow-up and case study are required to aid doctors in recognizing the range of MISC symptoms, which is required for prompt identification and suitable patient care.

Declaration by Authors

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