Acute Respiratory Distress Syndrome Complicating Pityriasis Rubra Pilaris

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Abstract
Pityriasis rubra pilaris is an uncommon inflammatory and hyperproliferative dermatosis of juvenile or adult onset. The etiology of the disease is still unknown. Acute respiratory distress syndrome has been described in generalized pustular psoriasis and/or erythrodermic psoriasis but it has never been reported in pityriasis rubra pilaris. We report a new case in a 73-year-old man with a 6 month history of pityriasis rubra pilaris. We review the published work on this complication when it has been associated with retinoids, pustular and erythrodermic psoriasis. The man developed acute respiratory distress syndrome and high-dose corticosteroid therapy was quickly initiated. Within a few days, his clinical and radiological status was dramatically improved. The pathogenesis of acute respiratory distress syndrome is unknown, but various proinflammatory cytokines have been implicated, especially tumor necrosis factor-alpha, which could play a role in the recruitment of leukocytes to the lung. This complication has never been reported but should be more widely known as the differential diagnoses include congestive heart failure, acute lung infection related or unrelated to immunosuppressive therapy, and drug hypersensitivity reaction. Early recognition would avoid delays in the correct management of this potentially lethal complication, which requires high-dose systemic corticosteroid therapy.

Keywords: Pityriasis rubra pilaris; Acute respiratory distress syndrome; Psoriasis; Retinoids

Introduction
Pityriasis Rubra Pilaris (PRP) is a heterogeneous inflammatory skin disease characterized by follicular papules, orange palmo-plantar keratoderma, and erythematous scaly patches with islands of skin sparing. Type I or classic adult PRP is the most common subtype and it usually goes with erythrodermia, a potentially life-threatening complication. It can result in fluid depletion, thermoregulatory compromise and maintained catabolic state. We present a patient who presented with PRP and six months later developed Acute Respiratory Distress Syndrome (ARDS) which is defined by the Berlin definition as a type of acute diffuse, inflammatory lung injury, leading to increased pulmonary vascular permeability, increased lung weight, and loss of aerated lung tissue [1].

Case Report
A 73-year-old man without medical history was diagnosed of classic PRP after developing progressive papulosquamous erythrodermia within one month. He had erythema affecting more than 95% of the skin and palmar keratoderma. Three incisional biopsies were taken showing alternating orthokeratosis and parakeratosis in both vertical and horizontal directions with focal hypergranulosis, thick suprapapillary plate, broad epidermal ridges, narrow dermal papillae and perivascular lymphocytic infiltrate in the superficial dermis and diagnosis of PRP was made (Figure 1A). A thorough study was performed including HIV testing, IGRA testing, tumoral markers and body TC and all the results were negative. He was started on oral acitretin 25 mg daily, increased to 50 mg after two weeks, clobetasol propionate 0.05% ointment and emollients. No improvement in his skin was observed after three months so the therapy was stopped.

Five days after stopping acitretin an unexpected deterioration occurred in his general condition. He became febrile (39°C) tachypnoeic and hypoxic with a neutrophil leukocytosis (14 × 10⁹ L). Chest X-ray showed widespread alveolar shadowing typical of ARDS (Figure 2). Flu test was negative. Two sets of blood cultures were negative, and a third grew Staphylococcus luddonensis.

Figure 1: A-Erythrodermia and palmo-plantar keratoderma; B-Patient on day seven after recovering from pulmonary symptoms.

Figure 2: Chest x-ray showing bilateral interstitial infiltrate.
Sputum and urine cultures were negative. Following consultation with the infectious diseases team Daptomycin and Ceftiraxone for infective endocarditis were commenced. Transesophageal echocardiography was normal. Surprisingly at this point, being treated only with antibiotics his skin spontaneously improved, remaining only a few papulosquamous lesions on the back and a slight palmo plantar keratoderma.

However, his fever and pulmonary status was sustained despite antibiotic therapy and he deteriorated further so he was referred to the intensive care unit requiring mechanical ventilation. At this point, respiratory failure was not fully explained by cardiac failure of fluid overload, ecocardiography was normal and his PaO2/FiO2 index was under 200. So a formal diagnosis of ARDS by the Berlin definition was made and he was commenced as a decision of the intensive care unit on bolus of intravenous methylprednisolone 200 mg daily for three days and progressive tapering. This decision was made when the suspicion of an autoimmune etiology was high.

At this point his fever settled and a rapid recovery of normal respiratory function was observed within five days. His recovery was sustained and radiological features had resolved at seven days. He was discharged home 15 days later on emollients and a reducing course of prednisone. He had no residual chest symptoms and his PRP was quiescent (Figure 1B). We could not draw a definite conclusion between the corticosteroid therapy and the improvement since it is the not the established treatment for ARDS or PRP [1]. No other microorganisms were isolated.

In this case the lack of response to antimicrobial agents initiated after culture results and following microbiology guidance suggests, although not confirms, that sepsis did not have a role in the development of ARDS, and no other microorganism were isolated. There is the possibility that both PRP and an infectious etiology did play a role in the development of ARDS as a pro-inflammatory stimulus. Although cases of acitretin related ARDS have been reported [2]. In these cases, the pulmonary symptoms are within the so called, Capillary Leak Syndrome (CLS), ATRA syndrome or differentiation syndrome which all appear to be the same pro-inflammatory reaction. In our case, the drug was also felt to be non contributory given that the clinical course was somewhat not compatible with a full developed CLS since the patient did not experience weight gain, edema, nor renal failure and acitretin had been stopped five days prior to the onset of ARDS which is inconsistent with the cases previously published. Given the timing of the development of ARDS it was felt most likely that the ARDS was a complication of PRP but it cannot be absolutely ruled out that acitretin played a role.

In psoriasis, pulmonary symptoms associated with generalized pustular psoriasis was postulated first in 1972 by Landry and Muller who described a case of ‘sterile pneumonitis’ [3]. There have been six case reports of ARDS complicating pustular psoriasis and three cases complicating erythrodermic psoriasis [4–10].

**Conclusion**

This case was of particular interest as life-threatening respiratory compromise secondary to ARDS developed and it led to spontaneous sustained skin recovery. Up to now we think this is the first case of ARDS complicating PRP.

**Conflict of Interest**

We declare that we do not have conflict of interest.

**References**