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A Case of Pityriasis Rubra Pilaris Associated with Myasthenia Gravis

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Abstract

Observation: Pityriasis rubra pilaris (PRP) is characterized by redness of the skin, and scaling with a variable degree of pruritus. It may rarely coexist with various neuromuscular diseases. There are many reports of multiple diseases such as pemphigus, vitiligo, alopecia areata, systemic lupus erythematosus, lichen planus, *Sjögren*'s syndrome, ulcerative colitis, *Hashimoto*'s thyroiditis, rheumatoid arthritis, lymphoma, and thymoma associated with myasthenia gravis (MG). We describe a patient with MG associated with PRP.

Introduction

PRP is an idiopathic papulosquamous disease [**1, 2, 3, 4, 5**]. The etiology of PRP is unknown [**2**]. PRP has been associated with *Down* syndrome, hyperthyroidism, leukemia, and myositis, as well as several autoimmune diseases including MG, hypothyroidism, celiac sprue, and vitiligo [**6**]. PRP association with neuromuscular diseases has been rarely reported. Herein, we describe a patient with MG associated with PRP.

Case Report

A 16-year-old female patient presented with symmetrical erythematous papules and plaques exhibiting a follicular pattern, involving dorsal and extensor surfaces of both lower and upper extremities, and abdomen in March 2001 (**Figure 1**). Physical examination was normal.

The result of routine complete blood cell count, urinalysis, tumor markers, chemistry group, thyroid hormones and erythrocyte sedimentation rate were within normal limits. Antinuclear and anti-DNA antibodies were negative, and IgG, A, M, total C3 and C4 complement levels were normal. HbsAg, anti-HbsAg, anti-HbcIgM, anti-HCV, HIV-

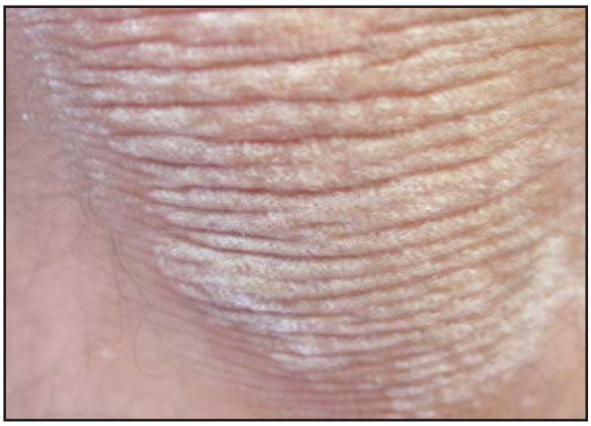

Figure 1. Erythematous papules and plaques exhibiting a follicular pattern, involving extensor surfaces of lower extremity

1, 2 antibodies were negative. A skin biopsy specimen obtained from the lower extremity of our patient revealed lamellar hyperkeratosis with focal parakeratosis and superficial perivascular lymphocytic infiltration, consistent with PRP. Based on clinical and histopathological findings, a diagnosis of PRP was made.

Our patient was treated with 200 000 international units of water miscible oral vitamin A daily (Avigen®, Eras, Turkey). The eruption resolved within few weeks and therapy was discontinued within three months. After 5 months, lesions recurred and systemic 20 mg daily isotretinoin treatment was started (Roaccutane®, Roche, Basel). Lesions regressed within 2 months and treatment was stopped. No relapse has been observed in a follow-up period of 6 months.

In November 2002, the patient developed a general fatigue, extraocular muscle weakness, dysarthria, and difficulty in climbing stairs, and complained of intermittent blurred vision, with greater severity of symptoms in the evening. In neurological examination, a diffuse symmetrical severe muscle weakness was most pronounced in the proximal muscles, but her tibialis anterior muscles were also involved bilaterally. The weakest muscles bilaterally were the deltoid, biceps, iliopsoas and tibialis anterior. There was no tenderness, myotonia, or atrophy. Repetitive stimulation test showed decrements of amplitude in ulnar nerve stimulation. Single fiber EMG of extensor digitorum communis muscle revealed increased neuromuscular jitter. Serum anti-Ach-receptor antibody was 65 nmol/ml (normal; 0-0.5nmol/ml). Her symptoms disappeared with an edrohoneum chloride test. The diagnosis of MG was made based on muscle weakness in the face and extremities, a positive edrohoneum chloride test, elevation of serum anti-Ach-R antibody titre and decrements of amplitude in a repetitive ulnar nerve stimulation test. She was administered 60 mg deflasocort on alternate days and pridostigmine 180 mg/daily. Her symptoms gradually improved.

Thymectomy was performed on March 2003, and thymus hyperplasia was detected on histopathological examination. On August 2003, she visited our department, and her myasthenic symptoms were almost recovered except for slight weakness of the lower limbs. Dermatological examination, revealed only a few red, scaling papules on the knees. We prescribed topical steroid for the lesions.

Discussion

PRP is an idiopathic papulosquamous disease first described in 1985 by *Claudius Tarral* [**1**]. The etiology of PRP is unknown. Although some researchers state that the development of PRP may be related to an abnormal immune response to antigenic triggers, the exact mechanism remains unclear [**2**]. PRP has been reported in patients with hypogammaglobulinemia and furunculosis, T-lymphocyte abnormalities, lymphocyte hypersensitivity to superantigens and HIV infection [**3, 4, 5**]. PRP has been associated with Down syndrome, hyperthyroidism, leukemia, and myositis, as well as several autoimmune diseases including MG, hypothyroidism, celiac sprue, and vitiligo [**6**]. MG is an autoimmune disease characterized by weakness of striated muscles resulting from the production of antibodies against acetycholine receptors in the neuromuscular junction. It is also associated with other autoimmune diseases such as pemphigus, vitiligo, alopecia areata, systemic lupus erythematosus, lichen planus, *Sjögren*'s syndrome, ulcerative colitis, *Hashimoto*'s thyroiditis, rheumatoid arthritis, hyperthyroidism and hypothyroidism [**7**].

MG associated with PRP is rarely reported in the literature. In 1950, *Kierland* and *Kulwin* found neuromuscular difficulties in 6 of 58 patients with PRP; one had myasthenia gravis, one had sclerodermatomyositis, and four had nonspecific generalized muscle weakness [**8**]. In 1965, *Waldorf* et al. described a case of vitamin A responsive PRP with MG [**9**]. The precise pathological mechanism of the association between PRP, thymus hyperplasia and autoimmune diseases are not fully understood. The thymus has been suggested to be a possible common origin of autoimmune response in this relationship. The thymus contains myoid cells and *Hassall*'s corpuscles, structures similar in appearance to muscle and epidermis, respectively. Myoid cells contain surface acetylcholine receptors. *Patten* et al. explain the etiology of this syndrome as follows: (i) abnormal immunoregulation occurs as a consequence of thymoma, thymic disease, or thymic insult; (ii) abnormal T-cell function leads to immune intolerance to squamous epithelium, cell nuclei, and striated muscle antigens

found in the *Hassall*'s corpuscle, and to myoid cells of the thymus resulting in production of autoantibodies to these structures; and (iii) ultimately, a cross-reactivity of these antibodies to normal skin and muscle antigen occurs resulting in an autoimmune disease [**10**]. Our cases suggest that exploiting a common genetic background focusing on the thymus could control antibodies in PRP and MG. To our knowledge, our patient is the third case of PRP associated with MG in the English literature. Considering this uncommon, but possible association, investigation of patients with PRP for a neurological disease is plausible.

References

1. Tarral C. General psoriasis. Desquamation from the parts covered with hair. In: Rayer PA. Theoretical and Practical Treatise on the Diseases of the skin, 2nd edn. London, Bailliere, 1835; 648-649.
2. Magro CM, Crowson AN. The clinical and histomorphological features of pityriasis rubra pilaris. A comparative analysis with psoriasis. J Cutan Pathol 1997; 24: 416-424. PMID: 9274959
3. Castanet J, Lacour JP, Perrin C, Brun P, Ortonne JP. Juvenile pitiyriasis rubra pilaris associated with hypogammaglobulinemia and furunculosis. Br J Dermatol 1994; 131: 717-719. PMID: 7999608
4. Yamamoto T, Yokoyama A. Lymphocyte response to superantigen in a patient with childhood-onset pityriasis rubra pilaris. Int J Dermatol 1999; 38: 639-640. PMID: 10487460
5. Blauvelt A, Nahass GT, Pardo RPJ, Kerdel FA. Pityriasis rubra pilaris and HIV infection. J Am Acad Dermatol 1991; 24: 703-705. PMID: 1869640
6. Behr FD, Bangert JL, Hansen RC. Atypical pityriasis rubra pilaris associated with arthropaty and osteoporosis: A case report with 15-year follow-up. Pediatr Dermatol 2002; 19: 46-51. PMID: 11860571
7. Goulon M, Estournet B, Tulliez M. Myasthenia Gravis and Associated Diseases. Int J Neurol 1980; 14: 61-72. PMID: 7348698
8. Kierland RR, Kulwin MH. Pityriasis rubra pilaris: A clinical study. Arch Dermatol 1950; 61: 925-930. PMID: 15419810
9. Waldorf DS, Hambrick GW. Vitamin A-Responsive Pityriasis Rubra Pilaris with Myasthenia Gravis. Arch Dermatol 1965; 92: 424-427. PMID: 5835336
10. Patten SF, Dijkstra JWE. Review associations of pemphigus and autoimmune disease with malignancy or thymoma. Int J Dermatol 1994; 33: 836-842. PMID: 7883404