Unusual association of diseases/symptoms

Arthritis associated with pityriasis rubra pilaris

Paul Y Liu,¹ Pamela E Prete²¹ Department of Medicine, Division of Rheumatology, University of California, Irvine, Orange, California, USA² Department of Medicine – Rheumatology, VA Healthcare System, Long Beach, California, USACorrespondence to Pamela E Prete, pamela.prete@va.gov**Summary**

Pityriasis rubra pilaris (PRP) is a rare papulosquamous disease of unknown aetiology, with an estimated prevalence of 2.5 per million.¹ It is clinically characterised by the presence of follicular hyperkeratotic papules, but the histopathology is usually non-specific. An interesting case of asymmetric, inflammatory arthritis of the hand in a patient who also presented with PRP is presented, along with a review of the few previously reported cases the literature.

BACKGROUND

Pityriasis rubra pilaris (PRP) is a rare skin disorder characterised by hyperkeratotic, papulosquamous lesions which can be associated with arthritis. Therefore, it is important for the clinician to be aware of such dermatologic associations when evaluating a patient presenting with musculoskeletal complaints. We present an interesting case of an asymmetric, inflammatory arthritis of the hand in a patient with PRP.

CASE PRESENTATION

A 63-year-old Caucasian male presented to rheumatology clinic with non-traumatic right hand and wrist pain and swelling. His symptoms had started insidiously and progressed gradually over 3 months. His primary care physician had prescribed ibuprofen 600 mg 3 times daily as well as a 2-week taper of high-dose oral prednisone, but there was no improvement. Concomitant with his right-hand

Figure 1 Photomicrograph of a skin biopsy specimen showing hyperparakeratosis, irregular acanthosis and chronic superficial perivascular inflammation compatible with pityriasis rubra pilaris. (Haematoxylin-eosin stain).

Figure 2 (A) Musculoskeletal exam before therapy. Soft tissue swelling and decreased flexion/extension in the right distal interphalangeals, proximal interphalangeals, MCPs and wrist. (B) Tri-phasic technetium bone scan demonstrating increased blood flow, blood pooling and delayed activity at the right wrist, and carpal bones and metacarpal-phalangeals consistent with inflammatory joint disease. (C) Musculoskeletal exam 6 months later. Pain and swelling have resolved and the range of motion is normal.

signs and symptoms, the patient developed a hyperkeratotic, follicular rash on the chest, back and extremities.

Significant in his past medical history was a diagnosis of PRP made 5 years earlier. At that time, he had multiple pruritic, scaling, well-demarcated, salmon-colored papules and plaques located on the scalp, face, neck, trunk, arms and legs. He also had waxy, thickened and erythematous lesions on his palms. Small erythematous papules were also noted on the dorsum of his hand and proximal phalanges. Periorbital edema was present, but nails and mucous membranes were normal. Two skin biopsies of the abdomen and back were performed, each revealing hyperparakeratosis and irregular acanthosis, compatible with PRP (figure 1). The patient was presented at Dermatology Grand Rounds, and a diagnosis of type 1 PRP was made. The patient was then treated with oral acitretin 0.5 mg/kg/day which resulted in a dramatic improvement in the rash within 4 weeks. After 3 months of treatment, the patient had only minimal erythema in the legs and hands; thus, acitretin was discontinued. The patient remained asymptomatic for 5 years.

INVESTIGATIONS

On presentation to rheumatology clinic, his musculoskeletal examination revealed soft tissue swelling and decreased flexion and extension in the distal interphalangeal, proximal interphalangeal, metacarpal-phalangeal joints and wrist (figure 2A). Pain was also elicited with active flexion and extension of these joints. Multiple-scale, erythematous, plaques were present on the chest, back and arms. Complete blood count, basic metabolic panel, liver function tests, serum protein electrophoresis and thyroid stimulating hormone were normal. C-reactive protein was 3.7 mg/l (normal 0.2–5), sedimentation rate was 26 mm/h and uric acid was 7.1 mg/dl. Rheumatoid factor, anticyclic citrullinated peptide antibody, hepatitis B surface antigen, hepatitis C antibody and HIV were negative. Antinuclear antibody (ANA) was positive with a titer of 1:320 in a nuclear pattern; antidouble-stranded DNA antibody was negative. Age-appropriate cancer-screening tests were also negative. A radiograph of the right hand showed periarticular soft tissue swelling of the right hand and wrist,

with mild degenerative joint changes in the radiocarpal, carpal and interphalangeal joints as well as multiple subchondral cysts in the lunate and distal ulna. A radiograph of the lumbosacral spine revealed degenerative disc disease without evidence of sacroiliitis. A triphasic technetium bone scan demonstrated increased blood flow, blood pooling and delayed activity in the right wrist, carpal bones and MCPs that were consistent with inflammatory joint disease (figure 2B).

TREATMENT

The patient was diagnosed with PRP-associated arthritis as no other etiology was found to explain his asymmetric inflammatory arthritis. His PRP was aggressively treated with topical steroids and his arthritis with anti-inflammatory drugs.

OUTCOME AND FOLLOW-UP

Over the next 4 months, both the patient's PRP and inflammatory arthritis improved with a marked decrease in pain and swelling. By 6 months, his symptoms completely resolved, and he regained full mobility and function in the right hand and wrist (figure 2C).

The patient was followed up 1 year after his initial presentation and remains in clinical remission.

DISCUSSION

PRP is an uncommon papulosquamous cutaneous disease of unknown etiology. It affects men and women equally and can occur in individuals of any ethnicity.² It is usually acquired, but inherited forms can also occur. Typical manifestations of type 1 PRP (classic adult type) include follicular

hyperkeratotic papules that can progress to become confluent areas of scaly, perifollicular erythema with patches of normal skin, referred to as 'islands of sparing'.¹ As in our patient, the palms and soles may also become hyperkeratotic with salmon-colored erythematous papules. Nail changes are also common and include yellowish discolorations, splinter hemorrhages and nail thickening. Although non-specific, common histologic findings include hyperkeratosis, parakeratosis, irregular acanthosis and dermal perivascular lymphocytic infiltrates as was noted in our patient.³

Although a few case reports of PRP-associated arthritis have been published, the relationship is not well established. The clinical features of PRP-associated arthritis as reported in the literature have been quite variable (table 1). Asymmetric (as in our case) or symmetric peripheral polyarthritis, axial disease and enthesitis have all been described. The hands, knees and wrists are the most common joints reported to be affected. Prior reports have described a variable temporal relationship between the onset of arthritis and PRP, in which arthritis may precede or follow the onset of PRP by months to years.³ In our patient, arthritis was diagnosed 5 years after the initial onset of PRP and developed simultaneously with a recurrence of his skin disease. Rheumatoid factor is negative in almost all of the reported cases as it was in our patient. Our patient also had a positive ANA as was recorded in two other cases of PRP with arthritis (table 1). Interestingly, a positive ANA has also been described in cases of PRP without arthritis.⁴ Only one case of erosive arthritis has been reported in association with PRP,⁵ though acro-osteolysis associated with nail changes has been reported multiple times. Our patient had neither acro-osteolysis nor erosive arthritis on repeat imaging.

Table 1 Reported cases of PRP-associated arthritis

Case reports	Year reported	Age (years)	Sex	Rheumatoid factor	Antinuclear antibody	HLA-B27	Arthritic site(s)	Arthritis treatment
Present case	2009	63	M	Negative	Positive 1:320	Unknown	Right hand, right wrist	NSAIDs*
Gül <i>et al</i> ⁸	2007	58	F	Negative	Negative	Positive	Lumbosacral	Methotrexate 10 mg/week
Chan <i>et al</i> ³	2004	50	M	Negative	Positive 1:160	Unknown	Right knee	Methotrexate
Nakafusa <i>et al</i> ⁹	2002	52	M	Positive	Negative	Negative	Neck, shoulders, elbows	Prednisolone 10 mg/day, bucillamine 100 mg/day
Behr <i>et al</i> ¹⁰	2002	12	M	Negative	Negative	Unknown	Hands, wrists	Etretinate 50–75 mg/day, methotrexate 15 mg/week, acitretin 25 mg/day
Conaghan <i>et al</i> ⁶	1999	17	M	Negative	Negative	Negative	Shoulders, knees, enthesitis	NSAIDs*, prednisolone, methotrexate, cyclosporin, TNF α receptor inhibitor, liarsazole
Fiallo <i>et al</i> ⁵	1996	19	M	Negative	Negative	Positive	Wrists, knees, ankles, sacro-iliac joint	Etretinate 0.5 mg/kg/day
Lister <i>et al</i> ¹¹	1989	21	F	Negative	Positive 1:160	Unknown	Neck, hands, knees, ankles	Methotrexate 10 mg/week
Duke <i>et al</i> ¹²	1987	18	M	Unknown	Unknown	Unknown	Hands	Unknown
Aguilar <i>et al</i> ¹³	1973	62	M	Unknown	Unknown	Unknown	Hands, feet	Chloroquine 250 mg bid, prednisolone 10 mg/day, aspirin

*NSAIDs

bid, twice daily; F, female; HLA, human leucocyte antigen; M, male; NSAIDs, non-steroidal anti-inflammatory drugs; PRP, pityriasis rubra pilaris.

Various pharmacologic therapies have been used to treat PRP-associated arthritis (table 1). Methotrexate, topical/systemic steroids, non-steroidal anti-inflammatory medications and retinoids have been used with varying success rates, in order to treat both PRP and PRP-associated arthritis. One case report describes the use of antitumour necrosis factor α (anti-TNF α) therapy in refractory, PRP-associated arthritis resulting in a dramatic improvement of the arthritis but not the skin.⁶ However, subsequent publications have described TNF α inhibitors to be effective therapeutic options for PRP.⁷ Our patient was not given methotrexate or a TNF α inhibitor as both conditions, PRP and inflammatory arthritis, resolved with a combination of topical steroids plus non-steroidal anti-inflammatory medications.

Although it is well known that dermatologic diseases such as psoriasis are associated with inflammatory arthritis, arthropathies can also be associated with rare dermatologic disorders such as PRP. It is important both diagnostically and therapeutically for the clinician to be aware of these skin associations when evaluating patients with musculoskeletal complaints.

Learning points

- ▶ Dermatologic diseases, such as PRP, can be associated with inflammatory arthritis. It is, therefore, important for clinicians to be aware of such associations when evaluating patients with musculoskeletal complaints.
- ▶ PRP-associated arthritis can have variable presentations, but appears to affect the hands, knees and wrists most often.
- ▶ PRP-associated arthritis is usually non-erosive.
- ▶ Optimal therapy for PRP-associated arthritis is not well defined; however, it may improve with adequate treatment of the cutaneous disease.

Competing interests None.

Patient consent Obtained.

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