

Images in Rheumatology

Relapsing Polychondritis With Palmoplantar Pustulosis

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Palmoplantar pustulosis (PPP) is a very rare cutaneous manifestation observed during relapsing polychondritis (RP),^{1,2} not found in other conditions associated with saddle nose (eg, granulomatosis with polyangiitis, sarcoidosis, VEXAS [vacuoles, E1 enzyme, X-linked, autoinflammatory, somatic] syndrome, congenital syphilis, leprosy, septal abscess).

A 56-year-old White woman presented with a 5-month history of progressive saddle nose deformity (Figure 1A) and recurrent nasal obstructions. There was no recent history of nasal trauma and no drug abuse. Examination revealed PPP (Figure 1B). Joint pain in hands and feet was reported for the past 10 years. C-reactive protein was < 5 mg/L. Antineutrophil cytoplasmic antibodies and syphilis serology (*Treponema pallidum* hemagglutination and venereal disease research laboratory test) were negative. We did not find a mutation in *UBA1* for VEXAS syndrome. A fluorodeoxyglucose positron emission tomography showed nasal cartilage, left sterno-costal cartilage, and thoracic aortitis hyperfixations. A diagnosis of RP was made. Erythematous basis and keratotic rims surrounding

palmoplantar lesions observed during syphilis can mimic PPP and systemic manifestations can also include syphilitic aortitis. No specific tests exist for RP diagnosis, and other more common conditions may need to be excluded before RP can be diagnosed, making its diagnosis a challenge for clinicians.³ Our patient was treated successfully with prednisone and oral methotrexate. At the 6-month follow-up visit, joint pain had resolved and PPP had disappeared.

REFERENCES

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Figure 1. (A) Saddle nose, and (B) palmoplantar pustulosis in our patient.