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Psoriatic onycho-pachydermo-periostitis

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VERSITA

Abstract

Psoriatic onycho-pachydermo-periostitis has been recognized as an uncommon subset of psoriatic arthritis, and to date, only a few cases have been reported. In general, psoriatic onycho-pachydermo-periostitis is regarded as a unique variant of psoriatic arthritis, but its pathology and pathophysiology are not well understood. Although psoriatic onycho-pachydermo-periostitis is usually found in patients with psoriasis, it can also be found in patients without psoriatic skin lesions. It is characterized by psoriatic nail changes (usually onycholysis), painful swelling of the soft tissue close to the distal phalanges, and radiographic changes of the distal phalanges with periosteal reaction and bone erosions. We present a 58-year-old man with a 3-year history of deformation, thickened nails and pustules on the skin of his fingers and toes, and painful redness of the nail bed accompanied with pain in small joints. The family history was negative. After confirmation of the diagnosis, methotrexate: 15 mg weekly, was initiated which led to symptoms improvement. Treatment of psoriatic onycho-pachydermo-periostitis is difficult. It is based on treatment modalities used for other forms of psoriatic arthritis, such as sulphasalazine, methotrexate, and anti-tumor necrosis factor antibody therapy with adalimumab and etanercept. Nonsteroidal anti-inflammatory drugs are usually ineffective. Retinoids, subungual cyclosporine and corticosteroid therapy also showed inefficient. In our patient, methotrexate has shown efficacy in symptom improvement.

Nail psoriasis is a common condition, particularly in patients with joint involvement. In psoriatic arthritis, distal interphalangeal joints are most frequently affected, but in some cases periphalangeal soft (connective) tissue thickening above the distal phalanges, including periosteal reaction, is present as well. Psoriatic arthritis has a significant negative impact on patient's quality of life, affecting physical activities, as well as emotional and social well being. Psoriatic onycho-pachydermo-periostitis is a recently described variant of psoriatic arthritis. It is characterized by psoriatic nail changes (usually onycholysis), painful swelling of the soft tissue close to the distal phalanges, and radiographic changes of the distal phalanges showing periosteal reaction and bone erosions (1). The disease is often refractory to treatment, and available therapeutic agents affect the nail-bed matrix with variable success.

Case report

We present a 58-year-old man, who was first admitted to our hospital in 2008, with a 3-year history of deformation, thickened nails and pustules on his fingers and toes, and recent painful redness of the nail bed, accompanied with pain in small joints. The family history was negative for similar conditions or skin disorders. Oral antifungal therapy was introduced in another institution, but without response.

Physical examination revealed tender, drumstick-like swelling of all fingers and toes. Bright-red erythema and scaling were observed on the distal area of the digits. The nail plates were partially destroyed with subungual hyperkeratosis (Figure 1 and 2). Arthralgia of the distal interphalangeal joints was present.

Complete blood count, electrolytes and liver-function tests were within normal limits.

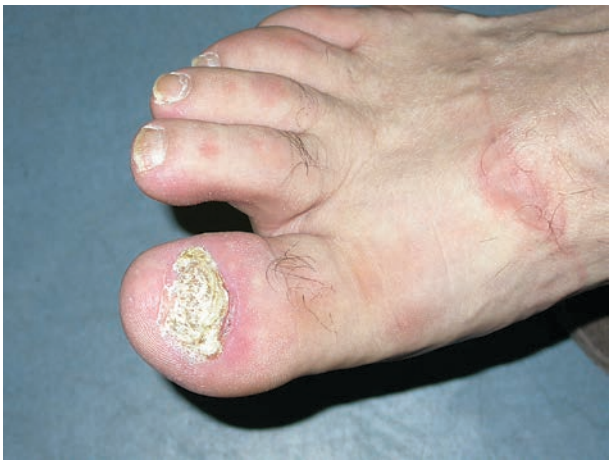

Figure 1. Swelling of the thumb

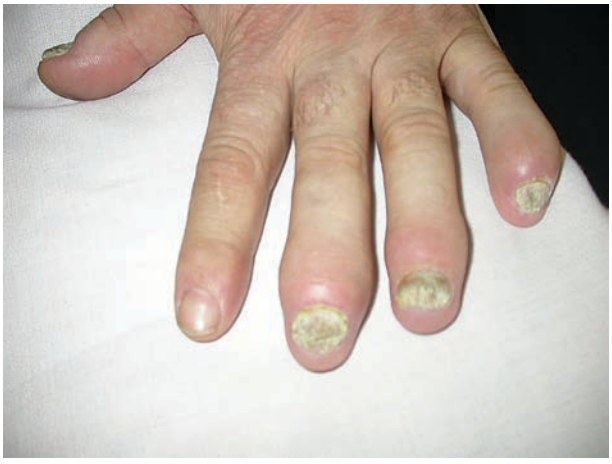

Figure 2. Swelling of the fingers (drumstick-like fingers)

The rheumatoid factor was negative, direct, native mycological examination and cultures were negative on three occasions. Nail bed biopsy was performed, and a skin sample was sent for histopathological analysis. It revealed parakeratosis and neutrophils forming Munro's abscesses in the corneal layer, features consistent with nail psoriasis (Figure 3). Direct immunofluorescence of skin sections was negative for IgG, IgM, IgA, C3 and fibrinogen deposits.

X-ray examination of hands, feet and sacroiliac joints revealed a preserved articular space width, but pronounced degenerative changes, especially in the distal interphalangeal joints with narrowing most prominent on the third finger of the left hand. Severe degenerative changes were also present on the distal interphalangeal joints of both thumbs, with preserved articular spaces. A considerable soft tissue swelling was also evident (Figure 4).

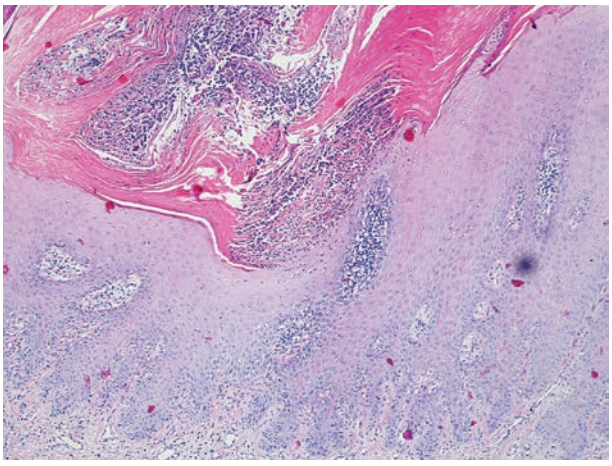

Figure 3. Histopathology analysis of the nail bed biopsy showing parakeratosis, intracorneal neutrophil pustule – Munro's abscess (Hematoxylin and Eosin, x 100)

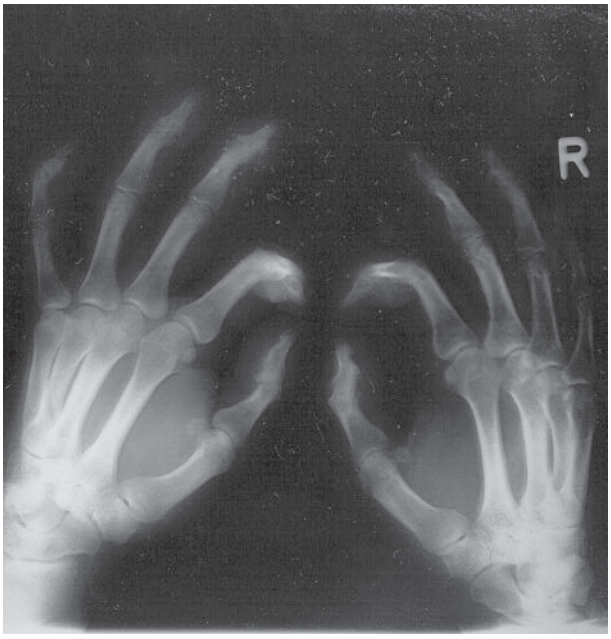

Figure 4. X-ray examination of hands revealing degeneration in the distal interphalangeal joints with narrowing most prominent on the third finger of the left hand, periostitis, swelling of the soft tissue

Based on the above-mentioned findings, the patient was diagnosed with psoriatic onycho-pachydermo-periostitis. After confirmation of the diagnosis, methotrexate therapy, 15 mg weekly, was initiated, which led to symptoms improvement.

The first control examination revealed a regression of erythema and desquamation on all fingers and toes. The pain was also significantly reduced. The treatment was continued with regular monitoring of the liver function. After three months of treatment, the pain was completely absent in the distal interphalangeal joints, and there were no new lesions on the nail plates. During the next six months, the dose was gradually reduced to 7.5 mg 1x a week, while maintaining good treatment outcome.

Discussion

Psoriatic onycho-pachydermo-periostitis was described by Fournier et all. in 1989 (1). It involves psoriatic onycho-distrophy and connective tissue thickening above the distal phalanges, including periosteal reaction. Psoriatic onycho-pachydermo-periostitis can be extremely painful. Thumbs are most commonly involved, but psoriatic onycho-pachydermo-periostitis may involve other digits as well.

Given the lack of distal joint involvement, but common nail involvement, bone changes in psoriatic onycho-pachydermo-periostitis are hypothesized to be caused by spread of inflammation from subungual dermis to the bone. The fibrous septum, which directly joins the subungual dermis and the distal phalanx and projects into the bone, may provide a route for the transmission of inflammation. Although psoriatic onycho-pachydermo-periostitis is usually seen in patients with psoriasis, it can also be found in patients without psoriatic skin lesions. It has been recognized as an uncommon subset of psoriatic arthritis and, to date, only a few cases have been described (1 - 4). In general, psoriatic onycho-pachydermo-periostitis is regarded as a unique variant of psoriatic arthritis, but its pathology and pathophysiology are not well understood.

The treatment of psoriatic onycho-pachydermo-periostitis is difficult. It is based on several treatment options, currently used for other forms of psoriatic arthritis, such as sulfasalazine, methotrexate, and anti-tumor necrosis factor antibody therapy with adalimumab and etanercept (3, 5). Nonsteroidal anti-inflammatory drugs are mostly ineffective. Retinoids, subungual cyclosporin and corticosteroid therapy are also inefficient. In our patient, methotrexate has shown efficacy regarding symptoms improvement.

Conclusion

Psoriatic onycho-pachydermo-periostitis is a recently described variant of psoriatic arthritis, which can be manifested in patients with or without other manifestations of psoriasis. Although there are no consistent data of satisfying therapy, and no guidelines in the treatment of this condition, all treatment options currently used for other forms of psoriatic arthritis may be of therapeutic benefit (3).

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Psoriatični onihopahidermoperiostitis

Sažetak

Uvod: Psorijaza često zahvata nokte, što se naročito često dešava kod artropatskog oblika psorijaze. U psorijatičnom artritisu najčešće su zahvaćeni distalni interfalangealni zglobovi, ali se u pojedinim slučajevima može javiti zadebljanje perifalangealnog mekog tkiva uz reakciju periosta. Psorijatični artritis u velikoj meri narušava kvalitet života obolelih, utičući na njihove fizičke, emocionalne i socijalne aktivnosti. Psorijatični onihopahidemoperiostitis je poseban oblik psorijatičnog artritisa, u kome postoje promene na noktima, najčešće oniholiza, bolno oticanje mekih tkiva oko distalnih falangi, uz prisustvo radiografski vidljivih promena na distalnim falangama u vidu periostne reakcije i erozija na kostima. Bolest je obično refrakterna na primenjene terapijske modalitete.

Prikaz slučaja: Prikazujemo redak oblik psorijatičnog onihopahidermoperiostitisa kod pedesetosmogodišnjeg muškarca. Prvi put se javio kod nas na pregled 2008. godine sa anamnestičkim podacima o trogodišnjem prisustvu deformisanih, zadebljalih noktiju, pojavi sitnih gnojnih plikova na prstima obe šake i stopala, crvenilu i bolnosti malih zglobova. Pacijent nije dao podatke o srodnicima obolelim od psorijaze. Antimikotični lekovi koje je pacijent uzimao peroralno u prethodnom periodu nisu doveli do poboljšanja.

Fizičkim pregledom pokazao je osetljivost, otok i izgled sličan palici na dobošu svih prstiju stopala i šaka. Distalni delovi prstiju bili su eritematozni i sa prisutnom deskvamacijom. Pored subungvalnih hiperkeratoza, nokatne ploče su pokazivale znake distrofije, a na nekim mestima su i nedostajale (slike 2 i 3). U distalnim interfalangealnim zglobovima šaka pacijent je osećao bol. Sve rutinske hematološke i biohemijske analize uključujući i reuma faktor bile su u granicama referentnih vrednosti. Uzorci izmenjenog tkiva u tri navrata su pregledani u nativnom mikroskopskom preparatu i u kulturi, ali nije potvrđeno prisustvo gljivične infekcije. Patohistološka analiza isečka uzetog sa nokatnog kreveca je pokazala promene koje se vide kod psorijaze noktiju, postojanje parakeratoze i neutrofilnih granulocita, koji su formirali u kornealnom sloju *Munro* apscese (Slika 2). Uzorci izmenjenog tkiva su pregledani u direktnom fluorescentnom mikroskopskom preparatu ali nije potvrđeno postojanje imunskih depozita.

Radiološki se na zglobovima šaka, stopala i sakroilijačnim zglobovma uočavala očuvana širina artikularnog prostora, uz vidljivo izražene degenerativne promene na distalnim interfalangealnim zglobovima sa suženjima naročito uočljivim na trećem prstu leve šake. Jako izražene degenerativne promene u distalnim interfalangealnim zglobovima uz očuvan artikularni prostor bile su prisutne na oba palca stopala, uz evidentan otok mekih tkiva oko distalnih falangi (Slika 3). Na osnovu gorenavedenih analiza, postavljena je dijagnoza psorijatičnog onihopahidermoperiostitisa. Pacijent je započeo lečenje metotreksatom u dozi od 15 mg nedeljno, nakon čega je usledilo kliničko poboljšanje.

Na prvom kontrolnom pregledu, uočena je regresija eritema i deskvamacije svim prstima šake i stopala. Osećaj bola je bio značajno smanjen. Lečenje je nastavljeno uz redovne laboratorijske kontrole funkcije jetre. Nakon tri meseca lečenja, osećaj bola je u potpunosti nestao na nivou distalnih interfalangealnih zglobova i nije dolazilo do pojave novih promena na nokatnim pločama. U toku narednih 6 meseci, doza leka je postepeno snižavana do 7,5 mg u jednoj dozi nedeljno uz očuvan dobar terapijski efekat.

Diskusija: Psorijatični onihopahidemoperiostitis prvi put je opisao 1989. godine Fournier sa saradnicima. Karakterišu ga promene na noktima, najčešće oniholiza, bolno oticanje mekih tkiva oko distalnih falangi i radiografski vidljiva periostna reakcija i erozije na kostima distalnih falangi.

Najčeće su zahvaćeni palčevi na stopalima, ali mogu biti zahvaćeni i svi ostali prsti stopala i šaka. Bolest je peaćena jakim osećajem bola, Iako se najčešće javlja kod obolelih od psorijaze, može da se javi i bez vidljivih psorijatičnih promena na koži. Oboljenje ima nedovoljno razjašnjenu etiologiju i patofiziologiju, ali smatra se da predstavlja poseban oblik psorijatičnog artritisa. Do sada je u svetskoj literaturi objavljen mali broj slučajeva. Pretpostavlja se da se zbog poštede distalnih interfalangealnih zglobova i postojanja promena na noktima, inflamacija sa subungvalnog dermisa preko fibroznih septuma prenosi na koštano tkivo. Bolest je obično tvrdokorna na primenjene terapijske modalitete koji deluju na matriks nokatnog kreveca, sa promenljivim terapijskim efektom, a koji se primenjuju za lečenje psorijatičnog artritisa: sulfasalazin, metotreksat, anti-tumor nekrozis faktor alfa antitela, adalimumab i etanercept. Lečenje retinoidima, subungvalno aplikovanim ciklosporinom, kortikosteroidima i nesteroidnim antiinflamatornim lekovima ne pokazuje željeni terapijski odgovor.

Zaključak: Prikazujemo slučaj retke terapijski tvrdokorne kliničke varijante psorijatičnog artritisa, u kome je primena metotreksata dovela do kliničkog poboljšanja.