**Table 1: Diagnostic criteria for APL**

Major criteria (4 out of 5 required for diagnosis)

1. Permanent and complete absence of scalp hair by the first few months of life
2. Few to widespread smooth, whitish, or milia-like papules on the face, scalp, arms, elbows, thighs or knees from infancy or childhood
3. Replacement of mature hair follicle structures by follicular cysts filled with cornified material in scalp histology
4. Mutation(s) in the human hairless gene through genetic testing
5. Clinical and/or molecular exclusion of vitamin D dependent rickets

Minor criteria (supplementary criteria)

1. Family history of consanguinity
2. Absence of secondary axillary, pubic, or body hair growth and/or sparse eyebrows and eyelashes
3. Normal growth and development, including normal bones, teeth, nails and sweating
4. Whitish-hypopigmented streaks on the scalp
5. Lack of response to any treatment modality

resistant rickets type IIA, a compound heterozygote for mutations in the Vitamin D receptor gene (VDR) in which the phenotype of atrichia with papular lesions was identical to that seen in patients carrying mutations in the HR gene. It is hypothesized that the VDR and HR genes, which are both zinc-finger proteins, may be in the same genetic pathway that controls postnatal cycling of the hair follicle.[4]

Yip *et al*.[5] proposed revised clinical criteria for APL based on their personal observation and have made a retrospective analysis of cases described in literature. These features are listed in Table 1.

Published estimates of the prevalence of APL remain surprisingly low, considering that pathogenetic mutations in HR have been found in distinct populations around the world.[2] APL is more common than previously thought and is often mistaken for the putative autoimmune form of alopecia universalis.[5]

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## REFERENCES

1. Indelman M, Bergman R, Lestringant GG, Peer G, Sprecher E. Compound heterozygosity for mutations in the hairless gene causes atrichia with papular lesions. Br J Dermatol 2003;148:553-7.
2. Online Mendelian Inheritance in Man, OMIM (TM). Johns Hopkins University, Baltimore, MD. MIM Number: {209500}. Available from: http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=209500 [last cited on 2003 May 9].
3. Ahmad W, Panteleyev AA, Christiano AM. The molecular basis of congenital atrichia in humans and mice: Mutations in the hairless gene (Symposium Proceedings/the Society for Investigative Dermatology). J Invest Dermatol 1999;4:240-3.
4. Miller J, Djabali K, Chen T, Liu Y, Ioffreda M, Lyle S, *et al*. Atrichia caused by mutations in the vitamin D receptor gene is a phenocopy of generalized atrichia caused by mutations in the hairless gene. J Invest Dermatol 2001;117:612-7.
5. Zlotogorski A, Panteleyev AA, Aita VM, Christiano AM. Clinical and molecular diagnostic criteria of congenital atrichia with papular lesions. J Invest Dermatol 2001;117:1662-5.

# Treatment of severe nail psoriasis with etanercept

Sir,

Nail involvement is common in psoriasis and has been reported to occur in up to 50% of patients.[1] It can cause significant physical impairment, severe distress and pain.[1,2] There is a broad spectrum of nail dystrophies associated with psoriasis, ranging from the common pitting and loosening of the nail plate to the less frequent discoloration and splinter hemorrhages seen in the nail bed.[1] It is frequently refractory to treatment and there is no standardized therapy regimen.[3] Etanercept is a fully humanized, soluble tumor necrosis factor (TNF)-alpha receptor approved for the treatment of plaque psoriasis. It has been shown to be safe and to have long-term efficacy for treatment of moderate to severe psoriasis resistant to other modes. Some reports show that etanercept may have significant benefit in the treatment of psoriatic nail disease, although not approved for its treatment (as only manifestation).[1-4]

We describe two cases of improvement of nail psoriasis with etanercept treatment. The severity of nail psoriasis was based on the Nail Psoriasis Severity Index (NAPSI).[5]

*Case 1*: A 59-year-old man with a 25-year history of plaque psoriasis, who after one episode of erythroderma and several systemic therapies (acitretin, methotrexate, cyclosporine), entered stabilization of the disease with Psoriasis Area Severity Index (PASI): 5 for the last 13 years. He was referred to our department because of severe nail psoriasis that was causing psychological distress. There was no improvement with topical application and intralesional injection of corticosteroids. All fingernails had psoriatic lesions (onycholysis, subungual hyperkeratosis, pitting and leukonychia) with NAPSI: 56. No symptoms of psoriatic arthritis were present. He was put on etanercept 50 mg twice a week (12 weeks), followed by etanercept 50 mg once a week (36 weeks), with NAPSI: 4 at week 48 (improvement of 92.9% of the nails' alterations) [Figure 1].

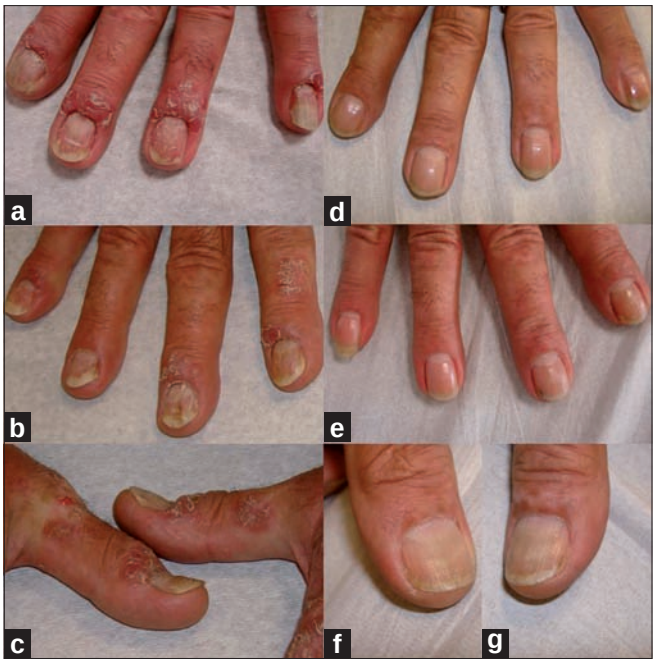


**Figure 1: (a–c) NAPSI: 56 at week 0 of treatment with etanercept; (d–g) NAPSI: 4 at week 48**

*Case 2*: A 43-year-old man with a 15-year history of plaque psoriasis recalcitrant to traditional systemic therapies was put on efalizumab, with significant clinical improvement of cutaneous psoriasis. After 20 months of efalizumab administration, he had PASI:1 but maintained onycholysis, subungual hyperkeratosis and pitting of all fingers (NAPSI: 53). No symptoms of psoriatic arthritis were noted. We decided to switch from efalizumab to etanercept 50 mg twice a week. At week 12, he was placed on a regimen of etanercept 25 mg twice a week. A significant improvement in nail psoriasis was noted after 1 year of etanercept treatment, with NAPSI: 30 at week 48 (56.6% improvement of the nail psoriasis) [Figure 2].

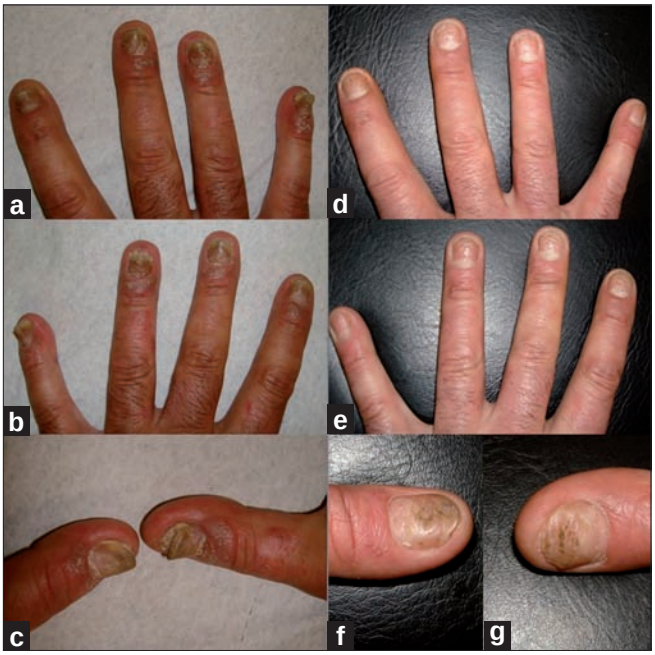


**Figure 2: (a–c) NAPSI: 53 at week 0 of treatment with etanercept; (d–g) NAPSI: 30 at week 48**

In both the patients, treatment was monitored closely. No side effects were present.

Recent studies have revealed that joint pain and nail psoriasis are responsible for impairment in quality of life. Etanercept therapy provided rapid improvement among patients with nail symptoms at baseline.[4]

In our two cases, the patients had been treated with several systemic therapies for psoriasis without improvement of nail manifestations. At week 48 of etanercept treatment, a good response was obtained with NAPSI improvement (92.9% and 56.6%, respectively). Both the patients were very much satisfied with the outcome.

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## REFERENCES

1. Jiaravuthisan M, Sasseville D, Vender RB, Murphy F, Muhn CY. Psoriasis of the nail: Anatomy, pathology, clinical presentation, and a review of the literature on therapy. J Am Acad Dermatol 2007;57:1-27.
2. Lawry M. Biological therapy and nail psoriasis. Dermatol Ther 2007;20:60-7.
3. Rallis E, Stavropoulou E, Rigopoulos D, Verros C. Rapid response of nail psoriasis to etanercept. J Rheumatol 2008;35:544-5.
4. Luger TA, Barker J, Lambert J, Yang S, Robertson D, Foehl J, *et al*. Sustained improvement in joint pain and nail symptoms with etanercept therapy in patients with moderate-to-severe psoriasis. J Eur Acad Dermatol Venereol 2009;23:896-904.
5. Rich P, Scher RK. Nail psoriasis severity index: A useful tool for evaluation of nail psoriasis. J Am Acad Dermatol 2003;49: 206-12.

# Dactylitis as a first manifestation of arthritis associated with hidradenitis suppurativa

Sir,

Dactylitis or "sausage digit" is a typical manifestation of spondyloarthropathy (SpA)[1] and it is so specific to SpA that it was included among the classification criteria of the whole disease group.[2] Dactylitis may sometimes occur alone for a long time as the only clinically manifestation of the SpA.[1]

We report here a patient with hidradenitis suppurativa (HS) in whom dactylitis occurred 2 years prior to the onset of a seronegative arthritis.

In October 2005, a 50-year-old Caucasian woman with a 10-year history of HS presented to the Dermatology Clinic with multiple pustules, nodules, abscesses, and cystic lesions in the axillary, mammary, and inguinal regions [Figure 1]. Furthermore, she complained of severe swelling and pain of the third finger of her left hand of 3 months duration. Her family history was negative for SpA and other HLA B27-associated diseases. There was no history of trauma, psoriasis, diarrhea, sexually transmitted diseases, conjunctivitis, iritis, uveitis, mucosal ulceration, Raynaud's phenomenon, sarcoidosis or gout. Clinical examination showed a swelling affecting the entire third finger of her left hand so pronounced that the patient could not flex it, and the presence of Heberden's nodules [Figure 2]. Laboratory studies showed an Erythrocyte Sedimentation Rate (ESR) of 101 mm/hour in the first hour, C-reactive protein (CRP) of 6.21 mg/dl, and a white blood cell count of 8.5 × 103/mm$^3$. Rheumatoid factor (RF), antinuclear (ANA) and anti-cyclic citrullinated peptide (anti-CCP) antibodies, circulating immune-complexes (CIC), angiotensin converting enzyme, HLA-B27 and HLA-DR4 antigens were normal or negative. Blood, urine and fecal cultures were negative, whilst

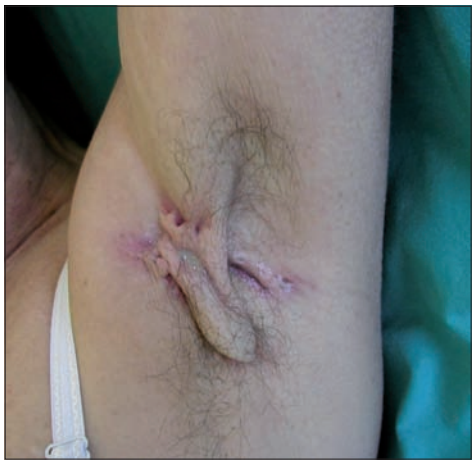

**Figure 1: Hidradenitis suppurativa: Evidence of nodules and cystic lesions in the left axillary region**

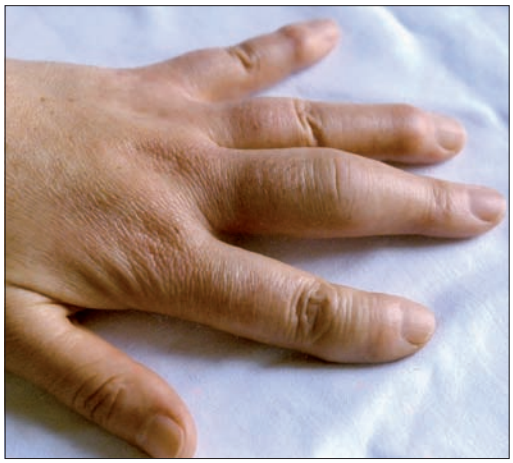

**Figure 2: Dactylitis: Evidence of severe swelling of the third finger of the left hand; as a collateral finding, we point out the presence of Heberden nodules on the second, fourth and fifth fingers**