Letters to the Editor

Sclerodermic lesions after liposuction in a patient with Raynaud’s phenomenon and anti-centromeric antibodies

Sirs,

Systemic sclerosis (SSc) is an autoimmune disorder of unknown aetiology. Trauma (1) and several pharmacological substances, including epinephrine (2, 3), have been identified as possible triggering pathogenic factors of this disorder. Here we describe the case of a patient with an overlooked prec scleroderma condition who developed scleroderma skin lesions after undergoing liposuction.

A 49-year-old Caucasian female was referred to us in June 1999, to evaluate the nature of extensive skin manifestations localised at the lower limbs at the sites of liposuction surgery performed in July 1998. The surgical procedure had been carried out using a solution of 0.05% lidocaine and 0.5 mg/ml epinephrine as local anaesthetic. The surgical trauma or the drugs might be responsible for the development of cutaneous lesions after episodes of physical trauma. J Rheumatol 1999; 26: 1816-7.


Treatment of TRAPS with etanercept: Use in pediatrics

Sirs,

Tumor necrosis factor-alpha receptor-associated periodic syndrome (TRAPS) is a recently described autosomal dominantly inherited disorder that manifests clinically as recurrent episodes of fever, erythematous rash, myalgia, arthralgia, abdominal pain and serositis (1). In contrast with familial Mediterranean fever (FMF), episodic attacks in TRAPS tend to be of longer duration, more responsive to glucocorticoids, and unresponsive to colchicine prophylaxis. The underlying defect in TRAPS is a mutation of the gene encoding the p55 TNF receptor (2). It is not yet clear whether excessive inflammation results from decrease-
ed soluble p55 TNF receptor (which may act as a TNF-alpha antagonist), changes in quantity or activity of the p55 receptor, shunting through the p75 TNF receptor, or some other mechanism.

Since TNF-alpha is central to the pathogenesis of TRAPS, this condition is potentially an excellent candidate for the new biologic TNF-alpha antagonists, etanercept and infliximab. Etanercept, which consists of two extracellular domains of the p75 TNF receptor fused to a human IgG1-Fc domain, is particularly appealing since it could potentially replace the role of soluble p55 TNF receptor. Early experience with etanercept in patients with TRAPS has been encouraging, and an open-label clinical trial is currently enrolling patients at the National Institutes of Health (1, 3).

Our patient is a 14-year-old female with episodes of localized inflammation since early infancy. Recurrent fever, rash and irritability led sequentially to the consideration of systemic-onset juvenile rheumatoid arthritis (JRA), familial Mediterranean fever and familial Hibernian fever. At age 11 she required laparotomy with resection of the cecum and terminal ileum for adhesions resulting from fibrosing serositis. Repeat surgery for lysis of adhesions was performed 2 years later. Empiric therapy with colchicine and prednisone was not tolerated. Partial improvement was obtained with oral tolmetin, but the patient still had monthly episodes of fever, arthralgia, cutaneous swelling or rash, and abdominal pain lasting one to three weeks. She was clinically diagnosed with TRAPS after her father, himself subject to similar recurrent fevers since age 11, was found to carry a T50M mutation in the p55 TNF receptor (2). Subsequent sequencing of the patient’s TNFRSF1A gene revealed the same mutation as her father. She has had no clinical evidence of amyloidosis. Aside from TRAPS and mild asthma, she has been well.

Treatment with etanercept was started at standard pediatric doses (0.4 mg/kg SQ twice weekly, maximum 25 mg). Over the 6 months since the beginning of the therapy, she has had no further episodes of fever or local inflammation, though she still experiences occasional arthralgia or myalgia. She reports that her sense of constant malaise has diminished, though she is still unable to exercise at the same intensity as her peers. Etanercept therapy has been without side effects.

Etanercept has rapidly become well accepted for use in pediatric rheumatic diseases. Early experience in JRA suggests a benign side effect profile (4), although the effects of chronic use in children are unknown. TRAPS is a rare disorder which leads not only to recurrent painful attacks of inflammation but also to long-term consequences, such as fibrosing serositis and in some cases amyloidosis. Our experience supports preliminary reports that etanercept may be effective and safe for children with TRAPS (1,5), though its long-term effects remain uncertain.

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References