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Recalcitrant pustular psoriasis responding to low dose Etanercept: a case report

Dermatology

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ABSTRACT

Generalized pustular psoriasis is a serious dermatological disease characterized by fever, chills and generalized pustulosis on the skin. Although, Etanercept is FDA approved biological for plaque type psoriasis, we gave a trial of this drug in a dose of 25mg/week for twelve doses with successful outcome. Case report: A 70 years old male with ten years history of chronic plaque psoriasis had one and half year's history of recurrent flares of pustular psoriasis. Histopathology confirmed the diagnosis of pustular psoriasis. Patient was treated with many systemic drugs including steroids, oral Acitretin, oral Cyclosporine and injection Methotrexate with no significant improvement. Considering unfavorable outcome with above drugs and adverse effects with long term steroids, patient was started on injection Etanercept 25 mg weekly with successful remission after twelve injections. Conclusion: We hereby report successful outcome while treating recalcitrant pustular psoriasis with low dose Etanercept.

Introduction: Generalized pustular psoriasis of Von Zumbusch is an unstable, inflammatory form of psoriasis, with the hallmark of neutrophil infiltration in cutaneous as well as extra cutaneous lesions. It is often recalcitrant, making treatment difficult. A slow and partial responses, as well as recurrences when the medication is decreased or suspended, are very frequent. The treatment of generalized pustular psoriasis has classically been carried out with oral Acitretin, Cyclosporine, Methotrexate and steroids. Biologicals like TNF alpha inhibitors have not been FDA approved for treating pustular psoriasis, although they are FDA approved for chronic plaque type psoriasis.

Case report: A70 years old male patient known case of chronic plaque psoriasis since ten years, presented with generalized pustulosis associated with fever and joint pains. There was history of fatigue and malaise.

On examination, patient had erythematous plaques with crops of pustules overlying it, over trunk, thighs and upper extremities. Patient was febrile, but his vitals were stable. Diagnosis was confirmed as pustular psoriasis on histopathological examination. Rest of the laboratory investigations were within normal limits. Ultrasonography abdomen and pelvis revealed mild fatty liver. Chest X ray, mantoux reading was done to rule out Koch's infection.

Patient was treated with intravenous steroids followed by Acitretin with no clinical remission of his flares of pustular psoriasis. In order to avoid side effects of systemic steroids on long term use and unsatisfactory response with Acitretin, consequently, patient was evaluated for Methotrexate and started on oral Methotrexate 15 mg weekly after stopping steroids and Acitretin after twelve months. Methotrexate was continued for twelve months, but still patient was having recurrent flares of pustular psoriasis. Due to mild fatty liver with hepatomegaly and abnormal liver function tests and also due to unsatisfactory effect of methotrexate, it was withheld and Cyclosporine [CysA] was introduced and continued for four months. During this four months period of treatment with CysA, patient's diseased state was still not under controlled. Patient was suffering from similar flares occurring every twenty to thirty days even though he was on CysA. Eventually, considering his lab and rising blood pressure parameters (not controlled by amlodipine), he was evaluated for injection Etanercept.

We then shifted the patient to injection Etanercept 25 mg per weekly subcutaneous dosing. Patient's flares were controlled in one month (4 injections) of giving injection Etanercept. Till date patient has received twelve injections at weekly interval without any undue side effects. Currently, patient is under total disease control since last six months.

Discussion: The etiopathogenesis of generalized pustular psoriasis includes production of chemokines, and the expression of adhesion molecules by keratinocytes and vascular endothelial cells that can be stimulated by TNF alpha produced within psoriatic lesions. These signals then cause recruitment of additional inflammatory cells into the lesion. TNF alpha may function as part of a positive feedback loop, which acts to amplify and sustain the inflammatory process within psoriatic lesions. Biologic responses that are induced or regulated by TNF are modulated by Etanercept. It may, therefore, serve to reduce inflammation by breaking this cycle (J.Kruegler, 2002).

Etanercept, is a dimeric, fully human fusion protein (produced in Chinese hamster ovary cell) consisting of two ligand binding domains of p75 TNFR fused to the Fc portion of human IgG. Etanercept binds to both soluble and membrane bound TNF alpha, thereby preventing the cytokines from binding to any cell surface receptor (Stephanie Mehlis, 2013). It has been shown to lessen disease severity in adult patients with psoriasis. Etanercept is FDA approved for chronic plaque psoriasis. However, there are few anecdotal reports of generalized pustular psoriasis responding to Etanercept (Esposito, 2008) (Fialová, 2014) (L.J.L., 2010).In our case, we also noticed complete resolution of lesion after twelve doses of injection Etanercept [25mg/week, subcutaneously].No undue adverse effects related to injection Etanercept were noted in our patient.

The standard dose of Etanercept approved for chronic plaque type psoriasis is 50mg/week for twelve doses. But, because of patient's financial constraints, we gave a trial of 25mg/week dose of Etanercept. To our surprise, our patient of generalized pustular psoriasis responded very well and is also in remission since past six months with suboptimal doses of injection Etanercept (25mg/week; 12 doses).

Conclusion: Etanercept, a TNF alpha antagonist biological was found to be useful for treating pustular psoriasis in our case. Its efficacy in treatment of pustular psoriasis requires more attention and proper studies in view of its FDA approval. As in our case, even suboptimal doses of Etanercept [25 mg/ week] can prove to be beneficial which can reduce the cost of the treatment.

Figure : 1 Crops of pustules present over trunk interspersed with multiple discrete scaly plaques

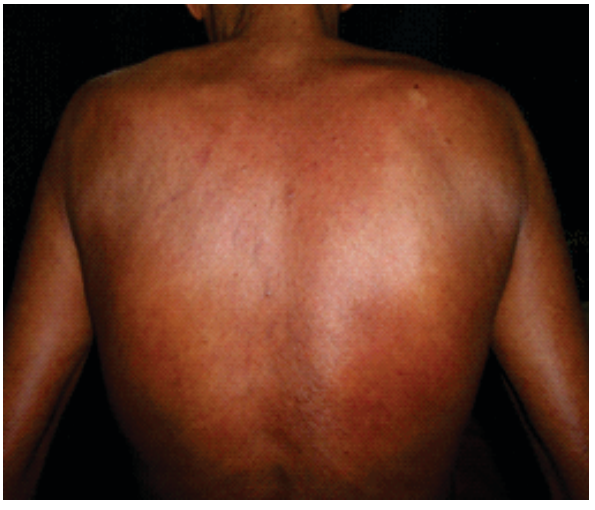

Figure: 2 Resolution of lesions after twelve injections of Etanercept

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