ABSTRACTS FROM CURRENT LITERATURE

Treatment of idiopathic nodular panniculitis (Pfiefer-Weber Christian Disease) with mycophenolate mofetil. Enk HA, Knop J. J Am Acad Dermatol 1998;39:508-509.

Idiopathic nodular panniculitis is characterised by fever and symmetric subcutaneous nodules often located on the lower extremities. In some patients there is involvement of the intraabdominal or retroperitoneal fat. Relapses are frequent and a fatal outcome has been described repeatedly. Therapy usually consists of steroids in medium to high doses as well as azathioprine or methotrexate. Even with this treatment the disease tends to relapse and has an uncertain prognosis.

Three febrile patients with relapsing nonsuppurating, multiple, painful subcutaneous nodules on the legs, arms and abdomen were used for the study and in them the diagnosis of Weber Christian disease was confirmed histologically. Two patients also had inflammatory lesions in their retroperitoneal fat as assessed by magnetic resonance imaging. All patients had a markedly elevated c-reactive protein, ESR and fever upto 40^0 C. No underlying illness was detectable. All patients were initially treated with 1.5mg /Kg body w.prednisolone.

Although 2 of the 3 patients showed marked improvement, attempts to lower the steroid dose resulted in prompt relapses. One patient showed no improvement. Therefore prednisolone in a dose of 2mg /Kg body weight was given to all patients, azathioprine in a dose of 1.5mg/Kg body weight to the initial responders and methotrexate in a dose 50mg/wk to the patient without initial response. Although all patients, responded, repeated attempts to reduce the steroid dose resulted in relapses. At this stage all 3 patients were given mycophenolate mofetil (2gm/day) in addition to prednisolone .

After 2 weeks the ESR and reactive protein had normalised and steroid doses were gradually discontinued without any relapse. All patients had no signs of disease activity during a 6-10 months follow up. Mycophenolate was reduced to 1gm/day in 2 of the patients without relapse. Only mild lymphopenia occurred in all patients. Mycophenolate mofetil is the 2 morpholinoethyl ester of mycophenolic acid. It is rapidly metabolised after oral administration to mycophenolic acid. Mycophenolic acid potently, selectively and reversibly inhibits inosine monophosphate dehydrogenase, a key enzyme in the de novo purine synthesis. Because lymphocytes primarily rely on the de novo synthesis of purines, mycophenolic acid acts as a strong immunosuppressive drug. It prevents T cell proliferation and inhibits B cells and antibody formation. Hence mycophenate is found to be highly effective in the treatment of relapsing panniculitis.

Pulse methyl prednisolone therapy for severe alopecia areata : An open prospective study of 45 patients. Friedli A, Labarthe MP, Engethardt E. J Am Acad Dermatol 1998; 39: 597-602.

Alopecia areata (AA) is an autoimmune disease of variable course with spontaneous remissions and frequent relapses. Available therapy includes topical, intralesional or systemic corticosteroids, PUVA, cryotherapy, topical sensitizers, anthralin, minoxidil and isoprinoline. To avoid the side effects of prolonged oral corticosteroids, pulse therapy was introduced by Burton and Shuster. In this study 45 patients with AA were se-

lected.

Among them 20 had multifocal AA, 10 had ophiasic AA and 15 had AA totalis and universalis. Selected patients bald area >30%, actual hair loss for <12 months, and all had clinically active disease. Each patient was given a single pulse of methyl prednisolone 250mg twice daily for 3 consecutive days. The rsult was assessed at 1,3,6, and 12 months after treatment and charted on a standardised sheet. Hair growth >20% at first month was considered as initial response to treatment and loss>25% during follow up as relapse. Multifocal alopecia showed best response. 50-100% response was present in 45%,60%, 65% and 60% at 1,3,6, and 12 months respectively. In ophiasic AA, there was no total regrowth at all. 40% showed 20-70% regrowth at 1 month with subsequent relapse at 3-6 months.

In the case of alopecia totalis and universalis, 53% had no response, 20% showed 50-90% regrowth at 1 month, 26% showed delayed response between 9 and 16 months, Among the 45 patients, 20 were of the first episode. Among them 14 responded well. The remaining 25 patients had relapsing alopecia. 17 of them did not respond to treatment. The following factors help in selection of patients: 1) first episode of alopecia showed good prognosis; 2) multifocal AA showed better response than ophiasic AA, totalis and universalis. Second pulse after some months is effective for relapses.

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Lichen sclerosus following the lines of Blaschko. Libow LF, Coots NV. J Am Acad Dermatol 1998;38: 831-833.

This is a case report of an unusual case of extragenital lichen sclerosus (LS) occurring along the line of Blaschko. A 25 -year- old white woman presented with a pruritic eruption over left lower side of the abdomen, left flank and left side of the back of 7 years duration. It was stable since 3 years and was not responding to medium to potent topical steroids and topical antibiotics.

Histopathology showed compact orthokeratosis and follicular plugging overlying a thinned epidermis devoid of rete ridges, papillary dermal edema and superficial and deep perivascular lymphocyto- histiocytic infiltrate with occasional plasma cells. Significant reduction in pruritus was observed with application of clobetasol cream,tetracycline (500mg twice daily) and topical tretinoin (0.05%), then clobetasol was gradually discontinued.

Extragenital lichen sclerosus is found in other parts of the body but LS which develops in a pattern corresponding to Blaschko's lines is rare. Disorders that occur along the lines of Blaschko are believed to result from two different clones of cells that evolve early in embryogenesis. Lyonization or random inactivation in X-linked disorders, postzygotic- somatic mutations in conditions and genetic half chromatid mutations have proposed as mechanisms.

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