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Clinical Image

Paraneoplastic Pemphigus in Setting of Waldenstrom's Macroglobulinemia

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Clinical Image

61-year-old male with history of treated Waldenstrom's macroglobulinemia presented with fever, sore throat and rash. Rash was characterized as pruritic, with confluent erythematous plaques and superficial pustules. Initially appeared on lower extremities and progressed to involve the entire body (Figure 1). Skin biopsy revealed a separated epidermis (subepidermal and suprabasilar clefting) with necrotic keratinocytes and eosinophils in the epidermis (Figure 1). Elevated serum antibody titers against Desmoglein 1 and 3, monkey-esophagus and rat bladder were noted. Direct immunofluorescence demonstrated intercellular staining for IgG with weak basement membrane staining for C3 suggesting a diagnosis of paraneoplastic pemphigus (Figure 1). Patient was then started on solumedrol, IVIG and rituximab with no improvement.

Patient subsequently developed multi organ failure requiring intubation, vasopressors, inotropic support, and hemodialysis before expiring. An autopsy showed atypical lymphoid infiltrate of the pancreas, liver, adrenal glands and lymph nodes consistent with involvement of Waldenstrom's macroglobulinemia.

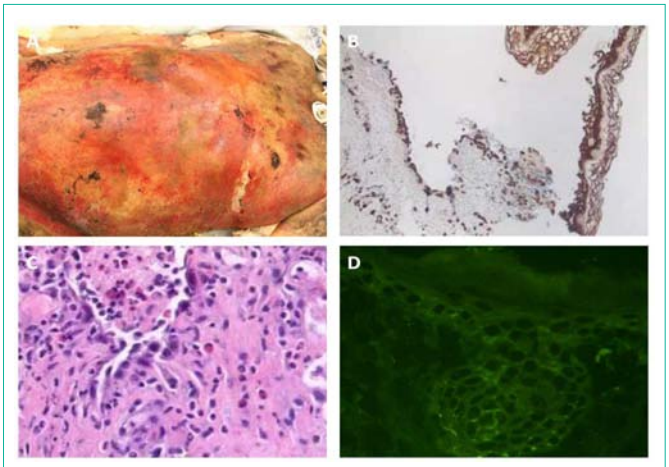

Figure 1: Panel A shows autopsy corpse with diffuse systemic dermatological findings consistent with paraneoplastic pemphigus secondary to Waldenstrom's Macroglobulinemia. Panel B and C shows scattered keratinocytic necrosis within the separated epidermis and a mixture of both subepidermal and suprabasilar clefting. Panel D showed the presence of serum cell surface IgG antibodies detected by cutaneous immunofluorescence testing on monkey esophagus substrate.

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