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MANTLE CELL LYMPHOMA PRESENTING AS BULLOUS PEMPHIGOID: A RARE PARANEOPLASTIC SYNDROME

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INTRODUCTION

- Mantle cell lymphoma (MCL) is a rare and aggressive hematological malignancy accounting for 5 to 7% of all lymphomas in the United States and Europe. Multiple paraneoplastic syndromes (PNS) have been described in MCL, ranging from neurological, endocrine, and autoimmune manifestations. There are two reported cases of bullous pemphigoid (BP) with MCL; we report the third case, adding to the evidence for this very rare association.

CASE DETAILS

- A 60-year-old male was admitted for a progressive blistering rash encompassing near-total skin surface, with rupture and hemorrhagic crusting with minimal external pressure for six days.
- Skin biopsy showed linear deposits of IgG and C3 along the epithelial basement membrane, consistent with BP. Treatment with topical and systemic steroids was instituted.
- The patient's white blood cell count was 14. 5 X 109 cells/L, and a peripheral blood smear to evaluate lymphocytosis showed immature and atypical lymphocytes.
- Peripheral blood flow cytometry and a core biopsy of the inguinal lymph node confirmed a diagnosis of mantle cell lymphoma, a pleomorphic variant.
- The patient started chemotherapy with a Bendamustine-Rituximab (BR) based regimen.
- Systemic and topical steroids were tapered off, and the patient remained in remission for BP with ongoing treatment for the MCL.

IMAGES



Images taken in the Emergency Department upon presentation of the patient. Images demonstrate widespread hemorrhagic bullae along with hemorrhagic crusting.

DISCUSSION

- BP is an autoimmune dermatologic condition prevalent in the elderly population, with reported associations to multiple solid organ tumors.
- The association between BP and hematological malignancies is very rare.
- Production of antibodies to tumor-specific antigens that might cross-react with the basal membrane zone has been postulated to explain the relationship.
- Alternate theories suggest that secondary production of anti-basement membrane zone antibodies by the cancer cells occurs through the secretion of a substance that damages the basement membrane.
- Controversies exist around the association or co-existence of BP with malignancies due to the increased prevalence of both in the elderly age group.
- The rarity of PNS in hematological malignancies makes establishing the association a challenge in less common cancers such as MCL.

CONCLUSIONS

- BP has rare associations with malignancies and has only been reported in the form of case reports or retrospective studies.
- Our case adds to the strength of evidence to study this association further in the form of large prospective studies.

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