



AUTOIMMUNE BULLOUS DISEASES

A CASE OF PARANEOPLASTIC PEMPHIGUS ASSOCIATED WITH LOCALIZED RETROPERITONEAL CASTLEMAN'S DISEASE

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Background: A 20-year-old female suffered with recalcitrant mucosal erosions and cutaneous lichen planus-like eruptions for half a year. She was diagnosed as pemphigus vulgaris (PV) and treated for 6 months with little improvement.

Observation: Level of Serum immunoglobulin G is 618 mg/dl and desmoglein (Dsg) 3 autoantibody is 185 U/ml. Ophthalmology examination revealed bitemporal retinal degeneration. Abdominal ultrasonography detected a solid mass at the lower pole of the left kidney. Then abdominal computed tomography revealed a heterogeneous 3.8x4.5 cm retroperitoneal solid lesion on the left of aorta with left hydronephrosis. Histologic examination of the oral lesion revealed subcutaneous blisters, suprabasal cleft formation, epidermal acantholysis, dermal papilla edema and lymphohistiocytic infiltration in the upper dermis, which were compatible with PV. Histologic examination of the cutaneous lesion showed epidermal hyperkeratosis, acanthosis, liquifaction degeneration of basal cells, serrated rete ridges and a dense band of inflammatory cells in the upper dermis, which were consistent with lichen planus. The clinical features and examinations suggest a diagnosis of paraneoplastic pemphigus (PNP). Furthermore the results of serum tumor markers revealed CEA 8.76 ng/ml, CYFRA 5.98 ng/ml, CA125 61.02 ng/ml, CA72-4 25.70 ng/ml. Indirect immunofluorescence disclosed a high titer of circulating immunoglobulin G (1:160) recognizing intercellular substances of rat bladder epithelium, also a specific screening test for PNP. After the intravenous administration of immunoglobulin, surgical resection was performed. The clinical diagnosis was PNP associated with Castleman's disease (CD), supported by the pathologic analysis. The patient had suffered from an exacerbation of the mucocutaneous lesions and severe bronchiolitis obliterans, and then a remittance within half a year. This case highlights the importance of PNP in patients with recalcitrant mucosal erosions, especially for young patients. We also suggest the importance of preoperative preparation and postoperative respiratory intervention for better prognosis.

Key message: pemphigus vulgaris, Paraneoplastic Pemphigus, Castleman's disease

