

Unusual distribution of bullous pemphigoid lesions in a hemiplegic patient: A case report

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Abstract Bullous pemphigoid (BP) is an autoimmune subepidermal blistering condition that predominantly affects older patients and characterized by local or generalized tense blister formation. Mucosal involvement with small blisters or erosions may exist in a minority of patients. BP is generally self-limiting with remission in most patients by 5 years. We report an 83-year-old hemiplegic lady with unusual distribution of bullous pemphigoid lesions.

Key words

Bullous pemphigoid, hemiplegia.

Introduction

Bullous pemphigoid (BP) is an autoimmune subepidermal blistering condition that predominantly affects older patients characterized by local or generalized tense blister formation. Mainly IgG autoantibodies bind to BP 230 and BP 180 antigens, components of the hemidesmosome adhesion complex, triggering activation of complement and release of tissue-destructive enzymes.¹ Mucosal involvement with small blisters or erosions may exist in a minority of patients. BP is generally self-limiting with remission in most patients by 5 years.

Case Report

An 83-year-old lady presented with a three-month history of developing bullous lesions on the skin. On examination she had multiple tense bullae and eroded areas. Most lesions were present on the flexor surface of left arm (**Figure 1**) with scattered lesions on left chest, left buttock and left leg. The lesions had a

strikingly unilateral distribution and no lesions were noted or reported on the right side of the body. No mucosal or ocular lesions were noticed.

The patient had a dense right-sided hemiplegia (**Figure 2**) and aphasia resulting from a left cerebrovascular accident (CVA) five years ago. The right arm had developed severe flexure contractures following the CVA. She had history of hypertension and was on aspirin 300 milligrams a day.

Skin biopsies were taken from appropriate site on the left arm and showed a subepidermal bulla and linear deposition of IgG and complement consistent with the diagnosis of BP. Biopsy was also taken from a corresponding site on right arm and was reported as normal histology and negative direct immunofluorescence. Indirect immunofluorescence did not demonstrate circulating anti-basement membrane autoantibodies in the serum.

She was treated with prednisolone 30mg a day and all the skin lesions cleared within 6 weeks, the steroid dose was tapered off to a maintenance dose of 7.5 milligrams per day along with potent topical steroid for any new

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Figure 1 Left-sided hemorrhagic bullae.



Figure 2 Right-sided hemiplegia.

developing lesions and the disease remains under excellent control.

Discussion

We report an unusual association between a bullous disease and a neurological disorder. This elderly lady who had hemiplegia on the right side of her body developed BP on her left side only.

Very few cases of unilateral BP with hemiplegia have been reported to date.¹ In these reported cases both these conditions were present on the same side. To our knowledge BP and hemiplegia occurring on the opposite sides of the body has not been reported previously.

Sparing of non-paretic limb had been described in literature, in many other skin disorders. Troilius reported five cases of eczema which mainly affected the normal, nonhemiplegic side.² Thomsen reported a patient who developed Beau's lines on the

fingernails of his non paretic side following generalized exfoliative dermatitis.³ Scleroderma sparing the hemiplegic side has been reported. Other skin diseases occurring on the hemiplegic side have been reported like seborrheic eczema and livedo reticularis.^{4,5} Medical conditions sparing the paretic limbs like rheumatoid arthritis, gout, osteoarthritis and scleroderma have been described in the literature.⁶⁻¹⁰

The exact nature of the causative factors that lead to the expression of bullous pemphigoid on the same or the opposite side of hemiplegia remain obscure. What exactly lead to this is not clear but certainly poses a number of queries. Why are some conditions bilateral? Has the nervous system got a role in causation or modifying this autoimmune condition? Might damage to the neuronal tissue prevent the development of bilateral disease? Might this be a hint to the etiopathogenesis of bullous pemphigoid and other similar conditions involving non-paretic limbs?

Decreased use of the paretic limb or a modification in its blood flow might be responsible for the sparing effect described above. There is no evidence that resting a neurologically intact limb prevents progression of bullous pemphigoid lesions. Decreased skin blood flow, as measured by phenolsulphonphthalein clearance time, has been noted in paretic limbs, but its pathophysiological significance is uncertain.¹¹ Modification of inflammation by neurological lesions has been shown experimentally. Section of the sciatic nerve before induction of adjuvant arthritis in rats causes sparing of the paretic limbs.¹² Autonomic dysfunction induced by the stroke may have had an ameliorating effect. What exactly causes bullous pemphigoid lesions on non-paretic side is still obscure and need further analysis and research. Logically, the condition at present is totally unexplainable.

Conclusion

We report this unusual case presenting with BP and hemiplegia on the opposite side of the body. It is very much likely, that in future, further reports of this mysterious protective effect of hemiplegia will be reported in literature. In depth evaluation of the factors operating in hemiplegic patient's modifying tissue reaction patterns may elaborate the pathogenesis of bullous pemphigoid.

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