A Case of Bullous Pemphigoid in a Patient with Dual Cancers, Fortuitous or Paraneoplastic?

Dear Editor,

Bullous pemphigoid (BP) is an autoimmune subepidermal blistering skin condition. It is characterised by the presence of immunoglobulin G (IgG) autoantibodies specific for the hemidesmosomal BP antigens BP230 (BPAg1) and BP 180 (BPAg2). Unlike pemphigus, it is not traditionally recognised as a paraneoplastic phenomenon. As BP becomes increasingly prevalent in our ageing population, it would be important to be on a lookout for paraneoplastic pemphigoid masquerading as BP.

Case Report

An 85-year-old Eurasian female presented with a 2-week history of blisters predominantly over the left ear, hands and thighs. She had a background of diabetes mellitus, ischaemic heart disease, mild renal impairment, gout and hypothyroidism. Further history revealed a loss of appetite and a weight loss of approximately 7 kg over a course of 3 to 4 months. Examination revealed tense intact bullae over the left ear, hands, thighs and haemorrhagic bullae on the buccal mucosa (Fig. 1a). There was no ocular involvement. In addition, a small firm lump measuring approximately 1.5 cm over the upper medial quadrant of the right breast was detected. There were no palpable abdominal masses or lymphadenopathy.

Full blood count showed normochromic normocytic anaemia with a haemoglobin of 10.7 g/dL (range, 11.5 to 15.0). Creatinine was raised at 290 umol/L (range, 50 to 90), consistent with her medical history of chronic renal failure. The liver function test was normal. Besides an elevated CA 19-9 of 41.9 U/ml (range, 0.0 to 30.0), other tumour markers tested such as CA 15-3, CA 125, CEA and alpha fetoprotein were normal.

Skin biopsy from the thigh showed a subepidermal blister associated with lymphocytic and eosinophilic infiltrate in the underlying dermis (Fig. 1b). Direct immunofluorescent showed linear deposition of IgG and C3 along the basement membrane. Indirect immunofluorescent to split skin substrate had a titre of >1/160 with a roof pattern. Ultrasound showed a spiculated 1.3 x 1.5 x 1.6 cm mass in the right upper inner quadrant of the right breast which was highly suspicious for malignancy. Histology of the right breast lump showed Scharff-Bloom-Richardson grade 1 infiltrating ductal carcinoma. This was estrogen receptor positive and...
Discussion

Paraneoplastic pemphigus is a better recognised entity than paraneoplastic BP. However, reports in Asia suggest a higher incidence of malignancies in BP patients compared to age-matched controls. In Japan, 5.8% of 1000 BP patients had malignancies. In China, this was of higher incidence of 6.7% of 104 BP patients. In Taiwan, there was an even higher incidence of malignancies in BP patients compared than paraneoplastic BP. However, reports in Asia suggest a higher incidence of malignancies in BP patients compared to age-matched controls. In Japan, 5.8% of 1000 BP patients had malignancies. In China, this was of higher incidence of 6.7% of 104 BP patients. In Taiwan, there was an even higher incidence of 15.1% of 86 patients with BP who had malignancies.1

Less than a handful of case reports have demonstrated a true paraneoplastic phenomenon with the presentation of breast carcinoma. One patient had radical mastectomy for an invasive ductal carcinoma of her right breast and a few months later presented with BP and bone metastasis.2 Another case report of 3 patients with neoplastic and mastopathic pemphigoid masquerading as BP.3

In our case, the blisters were only completely resolved after starting Tamoxifen and the patient remained blister-free even after prednisolone was ceased whilst only being on topical steroids. Even so, we recognise that we cannot be certain that the presence of BP with dual malignancies here was indeed a paraneoplastic phenomenon as the underlying tumours were not resected and yet the blisters resolved. Understandably, it can also be argued that BP has a higher incidence in an older age group, which naturally has a higher rate of undiagnosed malignancy. Nevertheless, as BP becomes increasingly prevalent in our ageing population, in addition to increasing reports of such a phenomenon, it will be necessary to stay vigilant to cases of paraneoplastic pemphigoid masquerading as BP.

Conclusion

In our case, the blisters were completely resolved after starting Tamoxifen and the patient remained blister-free even after prednisolone was ceased whilst only being on topical steroids. Even so, we recognise that we cannot be certain that the presence of BP with dual malignancies here was indeed a paraneoplastic phenomenon as the underlying tumours were not resected and yet the blisters resolved. Understandably, it can also be argued that BP has a higher incidence in an older age group, which naturally has a higher rate of undiagnosed malignancy. Nevertheless, as BP becomes increasingly prevalent in our ageing population, in addition to increasing reports of such a phenomenon, it will be necessary to stay vigilant to cases of paraneoplastic pemphigoid masquerading as BP.

REFERENCES


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Annals Academy of Medicine