WHEN BLISTERING DISEASE AND CUTANEOUS MALIGNANCY MEET: PARANEOPLASTIC BULLOUS PEMPHIGOID ASSOCIATED WITH SQUAMOUS CELL CARCINOMA

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An 86-year-old woman presented with two concomitant cutaneous conditions: a large occipital nodule progressively increasing in size (Figure 1) and a chronic pruritic eruption with excoriated lesions involving the back and upper limbs. The latter had initially been diagnosed as prurigo nodularis and treated with topical corticosteroids. Indirect immunofluorescence for circulating anti-skin antibodies was negative. While awaiting surgical excision of the occipital lesion, the patient developed worsening pruritus and tense blisters. Based on the clinical suspicion of bullous pemphigoid, subsequently confirmed by histopathology and direct immunofluorescence, a prolonged tapering course of systemic corticosteroids was prescribed, although the patient never started treatment. Remarkably, the blistering disease resolved completely after surgical removal of the occipital lesion. Histological examination identified the tumor as a keratinizing squamous cell carcinoma with trichilemmal differentiation, moderately differentiated and infiltrative, consistent with a malignant proliferating pilar tumor. Given the complete remission of bullous pemphigoid following tumor excision, in the absence of specific therapy or significant medication changes, a diagnosis of paraneoplastic bullous pemphigoid secondary to squamous cell carcinoma was made. This case highlights two clinically relevant aspects. First, according to current European guidelines (1), the diagnosis of bullous pemphigoid should not rely solely on indirect immunofluorescence because false-negative results may occur. Autoantibodies may target antigens other than BP180 and BP230 or epitopes not detected by standard assays. In our patient, serological testing was negative, whereas histology and direct immunofluorescence established the diagnosis. Second, although paraneoplastic autoimmune blistering diseases are more frequently associated with pemphigus, a neoplastic trigger should also be considered in selected cases of bullous pemphigoid. Previous reports have described an association between bullous pemphigoid and squamous cell carcinoma (2-4), suggesting that tumor-related antigens may induce an autoimmune response leading to blister formation (5, 6). This case further emphasizes the close relationship between neoplastic and inflammatory diseases, highlighting the importance of considering an underlying malignancy in atypical presentations of autoimmune blistering disorders.

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