Good's syndrome associated with multiple basal cell carcinomas

To the Editor: Wu and colleagues[1] present an interesting case report about a patient they consider to be affected by Good's syndrome accompanied by multiple large basal cell carcinomas (BCCs). This remarkable observation points to the potential problem that there are still no sufficiently precise criteria for the diagnosis of this rare disease. The consensus definition of Good's syndrome is thymoma with hypogammaglobulinaemia, a definition that is used in the available reviews, a large series, and multiple case reports.[2-5] In addition, there is often a CD4+ T cell lymphopenia. Consequently, the patients are prone to bacterial, viral and fungal infections, but they also frequently present with autoimmune manifestations, such as myasthenia gravis, pure red cell aplasia or lichen planus.[2-5]

The patient described by Wu and colleagues[1] had a thymoma, but, interestingly, he did not have hypogammaglobulinaemia, infections or autoimmunity. However, he displayed a large number of extended BCCs, which are not a classical complication of Good's syndrome. The response to the 23-valent pneumococcal polysaccharide vaccine was reported as "inadequate", without further details provided. The CD4+ T cell count was within the reference range, although with an inversion of the CD4+ to CD8+ ratio.[1]

Thus, in the absence of the defining criterion of hypogammaglobulinaemia (immunoglobulin M and A levels were normal), the question arises if this report really describes a new case of Good's syndrome, or rather a thymoma (without specifying whether benign or malignant) occurring together with BCCs. Maybe the patient is only at the beginning of the disease without fulfilling currently the two minimal consensus symptoms and signs, and hypogammaglobulinaemia, infections and/or autoimmunity will appear later. However, one should consider that this individual is already prophylactically treated with intravenous immunoglobulins.

Overall, the investigation of the immune system of this patient seems a bit limited to conclude on a direct link between an unclear and incompletely investigated immunodeficiency and the occurrence of a large number of BCCs. Moreover, the classification of this case as an atypical Good's syndrome is discussable. The follow-up of the patient might reveal valuable insight into the pathophysiology of the BCCs, with the hope that the current treatment might prevent him from developing a full spectrum immunodeficiency.

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