Good's syndrome associated with multiple basal cell carcinomas

In reply: We thank Zimmer and Ollert[1] for their insightful comments on our article.[2] We agree that the absence of a defining diagnostic criteria for Good's syndrome warrants a careful consideration of potential cases. Although our patient does not have the classical hypogammaglobulinaemia, autoimmunity or frequent infections, he does demonstrate other pertinent features reported in the literature.

The patient we describe has few B cells (2%; reference range, 7–27%) and inverted CD4+ to CD8+ T-cell ratio, both of which have been highlighted as significant features of Good's syndrome.[3] He also has an inadequate response to the 23-valent pneumococcal polysaccharide vaccine, which was evaluated through measuring his pre- and post-antibody titres. This vaccine activates only carbohydrate antigen determinants, which are the primary response antigens for B cells without T cell help. He showed a limited fourfold rise in antibody titres to Pneumovax (Merck Sharpe and Dohme) in six of the 14 serotypes (Box). This demonstrated antigen-specific immunodeficiency and met the criteria for intravenous immunoglobulin authorisation.

We agree that it is possible that our patient is in the early stages of the disease. In a small proportion of individuals, the diagnosis of thymoma has been found to precede the diagnosis of hypogammaglobulinaemia by one to six years;[4] therefore, it is possible that this deficit is still developing in this patient.[4,5] [Correction added on 17 April 2024, after first online publication: the second sentence has been revised.] Alternatively, Good's syndrome is a rare disorder and, perhaps, there is a spectrum of disease where not all criteria are present. Nevertheless, we do feel that the link to cutaneous malignancy is worth highlighting as an important possibility in the spectrum of this type of immunodeficiency.

Interestingly, short term follow-up of the patient seems to show clinical reduction in basal cell carcinomas (BCCs) with intravenous immunoglobulin. The patient received monthly intravenous immunoglobulin replacement between weeks 32 and 71. On follow-up in week 82, he was found to have developed only one further BCC in this treatment period. This may suggest a clinical response to intravenous immunoglobulin given the contrast to the speed and number of growing skin cancers he had before treatment. Regardless, long term follow-up of the patient will be valuable, and he continues to develop non-melanoma skin cancers but at a slower rate than on initial presentation.

Functional evaluation of humoral immunity with pre- and post-antibody titres to the 23-valent pneumococcal polysaccharide vaccine

Serotypes	Pre-immunisation antibody titres (µg/mL)	Post-immunisation antibody titres (µg/mL)	Response*
2	1.2	6.7	Adequate
4	< 0.1	0.3	Inadequate
6B	1.2	0.9	Inadequate
8	1.4	11.0	Adequate
9V	0.2	0.5	Inadequate
10A	1.0	1.8	Inadequate
11A	1.7	2.1	Inadequate
14	0.9	3.9	Adequate
15B	1.5	3.7	Inadequate
17F	0.2	1.4	Adequate
18C	0.6	1.0	Inadequate
19F	6.3	Unavailable†	na
20	0.8	1.5	Inadequate
23F	0.2	1.8	Adequate
33F	1.7	6.9	Adequate

na = not applicable. * An adequate response is a fourfold increase in antibody titre. † Unavailable due to technical issue. ◆ [Correction added on 17 April 2024, after first online publication: footnote has been added.]

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