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GOOD'S SYNDROME REVEALED BY RECURRENT SIGMOID DIVERTICULITIS

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ABSTRACT

Thymus is a primary lymphoid organ whose main function is the education of the T lymphocytes. Thymoma is uncommon neoplasia derived from epithelial cells of the thymus. Patients with thymoma frequently present with autoimmune disorders, mostly myasthenia gravis. Good's syndrome (GS) is an acquired adult-onset combined immunodeficiency typically characterised by the triad of thymoma, hypogammaglobulinaemia and increased susceptibility to recurrent infections. Immunoglobulin replacement is the main therapeutic strategy. Our knowledge of thymoma-associated immunodeficiency is predominantly derived from case reports or small series. We report the case of a patient with recurrent sigmoid diverticulitis that appeared after a thymectomy, who was subsequently found to have severe hypogammaglobulinaemia.

KEYWORDS

Good's syndrome, thymoma, hypogammaglobulinaemia

LEARNING POINTS

- Good's syndrome is a rare, acquired, adult-onset immunodeficiency.
- Good's syndrome remains an enigmatic disease.
- Immunoglobulin replacement is the main therapeutic strategy.

INTRODUCTION/BACKGROUND

In 1954, Robert Good described three patients with thymoma and hypogammaglobulinaemia[1]. Good's syndrome (GS) is a rare and unique immunodeficiency condition occurring especially in adults, who experience the triad of thymoma, hypogammaglobulinaemia and increased susceptibility to recurrent bacterial, viral and fungal infections. Patients are most commonly between the ages of 40 and 70 years and have a thymoma, low-to-absent B cells in the peripheral blood, hypogammaglobulinaemia and defects in cell-mediated immunity. Autoimmune diseases, mainly pure red cell aplasia, other haematological disorders and erosive lichen planus are a common finding. The pathogenesis of this immunodeficiency remains unclear and should be treated by resection of the thymoma and immunoglobulin replacement to maintain adequate IgG values.

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CASE DESCRIPTION

A 65-year-old man, who was previously healthy, was found to have a mediastinal mass in October 2023, which was discovered on a computed tomography (CT) scan. The mass was resected. Histopathological examination revealed a spindle cell thymoma, type AB, based on the World Health Organization classification. At this time, immunological screening was not done.

He had no symptoms consistent with myasthenia gravis and denied any history of ENT or respiratory infections. Since January 2023, he had reported five episodes of sigmoid diverticulitis and was treated by antibiotics. A colonoscopy was performed in December 2024 and confirmed chronic diverticulitis with a mild obstruction. In February 2025, he was admitted again for abdominal pain and fever linked to yet another episode of diverticulitis. *Campylobacter jejuni* septicaemia with transient neutropenia was diagnosed, and he was given azithromycin for ten days.

Because the recent history of thymoma, an immunodeficiency was suspected. Blood tests showed normal serum levels of haemoglobin, creatinine and calcium; serology of HIV was negative. Serum protein electrophoresis showed hypogammaglobulinaemia. No monoclonal gammopathy was seen on the blood protein immunoelectrophoresis, and screening of urine Bence-Jones protein was also negative.

The IgG serum level was 330 mg/dl (normal range 700–1500), IgA 28 mg/dl (normal range 100–600), IgM 10 mg/dl (normal range 22–240) and IgE <2 KU/l (normal range <114). Immunophenotyping lymphocyte analysis revealed a complete absence of B-lymphocyte, and T-lymphocyte subpopulations were in the normal range (TCD4+ 1,070 cells/mm^3 and TCD8+ 1,053 cells/mm^3). Testing for serologies vaccination showed non-protection for diphtheria and tetanus.

Intravenous immunoglobulins replacement therapy was started with the objective of an IgG serum level of about 600 mg/dl. Finally, regarding recurrent sigmoiditis, surgical treatment is under discussion with surgeons.

DISCUSSION

In 1954, Robert Good described three patients with thymoma and hypogammaglobulinaemia[1]. Good's syndrome is a rare and unique immunodeficiency condition occurring especially in adults, who experience the triad of thymoma and hypogammaglobulinaemia, and increased susceptibility to recurrent bacterial, viral and fungal infections. Our patient denied any history of ENT and respiratory infections; he suffered only recurrent sigmoiditis. He presented hypogammaglobulinaemia and absent B cells in the peripheral blood. Moreover, no CD4 T cell deficiency was observed in this case.

Our knowledge of thymoma-associated immunodeficiency is predominantly derived from case reports or small series. GS occurs in 3–4% of patients with thymoma. In a systematic review of 132 patients with GS by Kelesidis and Yang, the diagnosis of thymoma preceded the diagnosis of hypogammaglobulinaemia, infection or diarrhoea in 42% of patients with an interval of 3 months to 18 years[2]. Zaman and al. observed no improvement in the hypogammaglobulinaemia in a large series of Good's syndrome[3].

The general consensus was that GS has a worse prognosis than other humoral immunodeficiencies. In a recent study, physicians of GS patients completed two questionnaires with a two-year interval, using data on 47 patients – 499 patient years in total. Results on epidemiology, disease characteristics and outcome are presented. Mean age at diagnosis was 60 years and median follow-up from onset of symptoms was 9 years; there was a high frequency of respiratory tract infections due to encapsulated bacteria; median survival was 14 years. Survival was reduced compared to age-matched population controls[4].

Some controversies exist as to whether this is a unique disease, or a subgroup of common variable immune deficiency (CVID). Recently, some distinguishing aspects of both syndromes have come to light reflecting fundamental differences in their underlying pathophysiology. GS and CVID differ in demographic features and immune phenotype. GS is found almost exclusively in adults and is characterised by a significantly reduced or absence of peripheral B cells. In CVID, which also occurs in children, most patients have normal or slightly reduced peripheral B cells, with a distinguishing feature of low memory B cells[4-8].

To conclude, Good's syndrome remains an enigmatic adult immunodeficiency whose only treatment is immunoglobulin replacement.

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