An Autopsy Case of Good's Syndrome Diagnosed Based on the Presence of Thymoma

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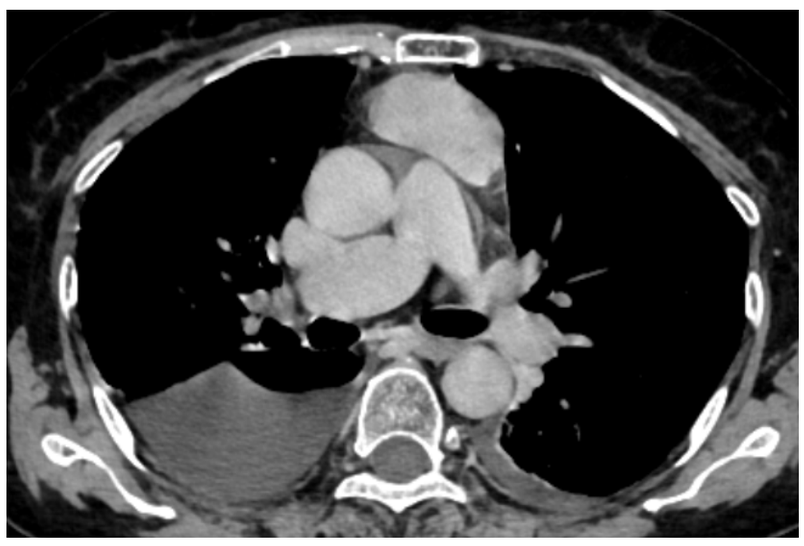
BACKGROUND

Good's syndrome is a rare adult-onset immunodeficiency characterized by thymoma, hypogammaglobulinemia, B-cell depletion, and T-cell abnormalities. Due to its rarity, diagnosis is often delayed.

CASE PRESENTATION

A 73-year-old woman presented with persistent fever. Despite empirical antibiotic therapy, her symptoms did not improve. Imaging studies revealed ascites and pleural effusion, and she was referred to our hospital. The ascites was exudative, but cytological examination was negative, consistent with infectious ascites. She was diagnosed with refractory bacterial peritonitis, and diagnostic laparoscopy was performed. However, no gastrointestinal perforation or other definitive infectious source was identified. Multiple antimicrobial regimens were administered, but her condition showed little improvement. Her medical history included thymoma. Immunological testing on day 15 showed severe hypogammaglobulinemia (IgG 186 mg/dL, IgA 20 mg/dL, IgM 3 mg/dL), and flow cytometry revealed a marked reduction of B cells, suggestive of Good's syndrome. Intravenous immunoglobulin (IVIG) therapy was initiated. However, her condition rapidly deteriorated, and she died on hospital day 20. Postmortem examination showed no specific infectious or neoplastic findings but demonstrated diffuse inflammatory changes. The clinical course was consistent with Good's syndrome.



Ascites biochemical test

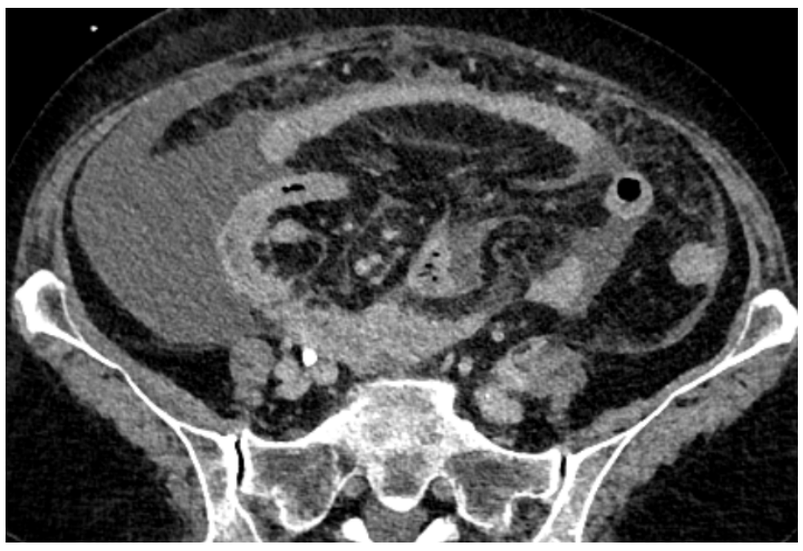
TP	3.0	g/dL
Alb	1.6	g/dL
LDH	3156	U/L
Glu	25	mg/dL
TNC	4904	/μL
Neut%	75.0	%
SAAG	0.5	g/dL

Immunological testing

IgG	186.0	mg/dL
IgA	20.0	mg/dL
IgM	3.0	mg/dL
C3	143	mg/dL
C4	29.4	mg/dL
CH50	>60.0	U/mL

Flow cytometry

CD4/CD8	0.4	
CD4	29.3	%
CD8	68.7	%
T cell	94.6	%
B cell	0.0	%



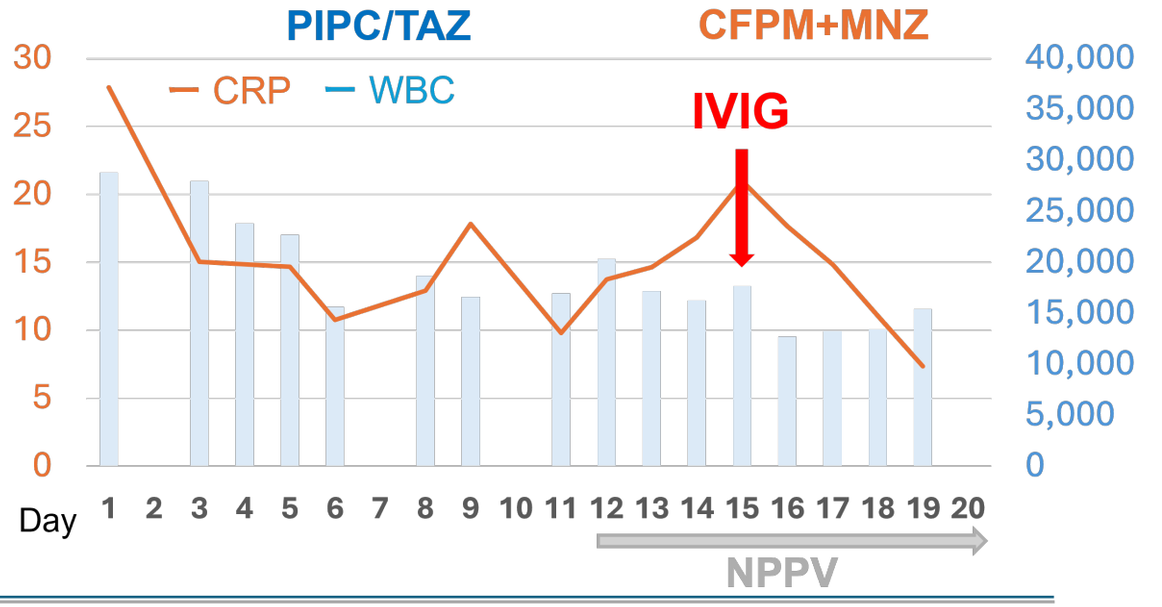
Ascities cytology

Negative

Microbiological cultures

Blood	No growth
Ascities	No growth

CT scan on day 1 showing ascites and pleural effusion, with a thymoma



DISCUSSION

Good's syndrome is primarily characterized by thymoma-associated immune deficiency and consequent infections, typically occurring in individuals aged 40–70 years. It is a very rare adult-onset disease, with fewer than 400 reported cases worldwide to date, mainly from Europe and Asia. The prevalence is estimated at 1 in 500,000–700,000. Both genders are equally affected, and no familial clustering has been reported. The pathogenesis remains poorly understood. No specific treatment protocols exist; the mainstay of therapy is regular immunoglobulin replacement combined with effective management of individual infections. Surgical intervention with complete thymectomy is clearly justified, even though it neither reverses the immune abnormalities nor prevents recurrent infections. In the present case, the recognition of Good's syndrome was delayed, leading to a postponement of immunoglobulin replacement therapy. Infection control proved difficult, and surgical intervention could not be performed. The patient died on hospital day 20. Earlier recognition of the syndrome might have led to a better outcome.

CONCLUSION

Although rare, Good's syndrome should be considered in adult patients with unexplained persistent infections and a history of thymoma. Early recognition and prompt initiation of immunoglobulin replacement therapy may be essential to improve outcomes. It is also suggested that the degree of immunodeficiency may be more severe than the level of hypogammaglobulinemia alone would indicate, and further accumulation of case reports is warranted.

REFERENCE

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3) Malphettes M, et al, Clin Infect Dis. 2015; 61; e13-e19