CASE REPORT

Good’s Syndrome Accompanied by Agranulocytosis Following a Rapid Clinical Course

Takahiro Okusu, Taiki Sato, Yoshitaka Ogata, Shinpei Nagata, Kazuhiro Kozumi, Sung-Ho Kim, Suguru Yamamoto and Shigeru Yamayoshi

Abstract

Good’s syndrome is an immunodeficiency disease involving thymoma accompanied by hypogammaglobulinemia. We encountered a case of Good’s syndrome accompanied by agranulocytosis that followed a rapid clinical course. A 72-year-old man visited our hospital with a two-week history of a sore throat. Candida albicans was detected in the pharynx, and hypogammaglobulinemia was detected in addition to granulocytopenia. The patient subsequently developed septic shock and followed a rapid clinical course which ended in death. Good’s syndrome with agranulocytosis was diagnosed at autopsy. Good’s syndrome accompanied by agranulocytosis can follow a rapid clinical course and some cases remain asymptomatic until old age. Its prompt treatment is crucial.

Key words: Good’s syndrome, agranulocytosis, hypogammaglobulinemia, thymoma, septic shock, autopsy

(Intern Med 55: 537-540, 2016)
(DOI: 10.2169/internalmedicine.55.5542)

Introduction

Good’s syndrome was first reported in 1955 by Robert Good as an immunodeficiency disease involving thymoma accompanied by hypogammaglobulinemia. Cases that also show agranulocytosis are rare (1). We encountered a patient with Good’s syndrome whose condition rapidly deteriorated following the development of a complication of agranulocytosis. The diagnosis and treatment of the patient proved difficult because his symptoms first appeared in old age.

Case Report

The patient was a 72-year-old man who had previously been healthy. The patient had no anamnesis or family medical history of note, and no history of smoking or drinking alcohol. The patient visited our hospital on an outpatient basis with a 2-week history of a persistent sore throat. A physical examination showed the following: body height, 150 cm; body weight, 56.6 kg; blood pressure, 151/109 mmHg; heart rate, 82 beats/min; and body temperature, 38.6°C. Extensive white moss was observed in the pharynx and on the tongue.

An initial full blood examination showed a hemoglobin level of 14.1 g/dL, a platelet count of 411×10^9/L, and a leukocyte count of 300/μL (1% neutrophils; 0% eosinophils; 4% basophils; 90% lymphocytes, 5% monocytes). The patient’s immunoglobulin (Ig) levels were as follows: IgG (493 mg/dL), IgA (50 mg/dL) and IgM (<16.9 mg/dL).

Computed tomography (CT) revealed a 63 mm×40 mm, well-circumscribed mass of non-uniform density in the anterior mediastinum, but no abnormalities were noted in the lung fields (Fig. 1). Candida albicans was detected in the pharynx, and Pseudomonas aeruginosa was isolated from a sputum culture. A bone marrow puncture showed hypoplasia of only leukocyte progenitor cells, and the patient experienced repeated bouts of septicemia despite the administration of antibiotic therapy (Fig. 2).

Complete atrioventricular block developed suddenly on day 13. The patient’s serum creatine kinase level became elevated (0.662 ng/mL, 2,545 U/L; normal range: 0 to 0.1 ng/mL, 30-200 U/L; no significant stenosis of the coronary arteries was found, and left ventricular angiography showed decreased wall motion of the entire circumference. Myocarditis was thus diagnosed. Artificial respiratory manage-
ment and circulatory support using catecholamines were in-
itiated, and the administration of antibacterial drugs and in-
travenous immunoglobulin were continued; however, the pa-
tient died on day 26.
At autopsy, the bone marrow showed hypopcellularity, the
myeloblasts were nearly arrested and mature granulocytes,
normal erythroid maturation and megakaryocyte lineages
were completely absent. There were no dysplastic features
noted. The tumor in the anterior mediastinum was identified
as a thymoma (World Health Organization type B2) (Fig. 3).
The myocardium showed extensive infiltration by lympho-
cytes and histiocytes as well as degeneration and destruc-

Figure 1. Computed tomography (CT) revealed a 63mm×40mm, well-circumscribed mass of non-
uniform density in the anterior mediastinum.

Figure 2. Bone marrow puncture showed hypoplasia of only leukocyte progenitor cells.

Figure 3. The tumor is a 63mm×40mm, well-circumscribed mass in the anterior mediastinum, and
it was identified as thymoma (World Health Organization type B2).
Good’s syndrome in patients who remain asymptomatic until old age

Reviews of the Good’s syndrome cases that have occurred to date show that the mean age at diagnosis is 59.1 years. Many cases are first diagnosed in middle age or later. At the time of the diagnosis of thymoma, 42.4% of cases are asymptomatic (10). However, the 5-year survival rate from the time of diagnosis is 70% and the prognosis is poor, with a majority of patients dying of infection, irrespective of the length of the asymptomatic course (11, 12).

Immunodeficiency in elderly patients is most often secondary immunodeficiency, caused by malignant tumors, infectious diseases, malnutrition, metabolic diseases, and drugs (13). Care must therefore be taken in cases (such as patient of the present study), in whom primary immunodeficiency develops suddenly in old age, when it can be easily overlooked. (14).

Cases of Good’s syndrome can be accompanied by agranulocytosis and may follow a rapid clinical course, however, patients can also remain asymptomatic until old age. Good’s syndrome accompanied by agranulocytosis follows a rapid clinical course after the development of severe immunodeficiency, and prompt treatment is crucial.

The authors state that they have no Conflict of Interest (COI).

References


© 2016 The Japanese Society of Internal Medicine
http://www.naika.or.jp/imonline/index.html