

## *Strongyloides* Hyperinfection in a Patient with Thymoma - A Rare Presentation of the Good Syndrome

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### Abstract

We present a case of *Strongyloides stercoralis* hyperinfection syndrome in the frame of a Good syndrome. The Good Syndrome is a rare cause of combined B- and T-cell immunodeficiency that occurs in association with a thymoma. Patients affected with Good syndrome have increased susceptibility to infections.

**Keywords:** *Strongyloides stercoralis*; *Strongyloides* hyperinfection syndrome; Good syndrome; Thymoma

### Case Report

We present the case of a 53-year old male from the Democratic Republic of Congo (DRC) who presented himself to his general practitioner with a long-lasting infection of the upper airways that did not respond to antibiotic therapy. A chest X-ray was performed and the patient was hospitalized because of a mediastinal mass. After CT-scan and a biopsy the diagnosis of a Thymoma Type AB Masaoka Stadium II was established. The surgical in toto thymectomy was performed without complications. A few days postinterventionally the patient presented with serious alteration of his general condition, constipation, vomiting after food intake and weight loss to now 35 kg, representing a BMI of 13.7 kg/m<sup>2</sup>. Symptomatic therapy was initiated and the patient was discharged. The patient was seen again by the thoracic surgeons for a follow up and removal of the surgical sutures. Because of a further alteration of his general condition and persistent gastrointestinal symptoms the patient was admitted to our clinic.

We performed a CT-scan of thorax and abdomen, diagnosing a nosocomial pneumonia consistent with low grade fever and a cough. We initiated calculated antibiotic therapy. Laboratory results revealed elevated CRP, normal leucocytes, no eosinophilia and most interestingly an immunodeficiency both linked to a low T-cell and B-cell count. A HIV-test showed negative (Table 1). Due to the persistence of the gastrointestinal symptoms we performed a gastroscopy. Histologically we were able to show larvae of nematodes-suspected to be *Strongyloides stercoralis*.

Further laboratory examinations showed negative HTLV-1 antibodies and a hypogammaglobulinemia of 2.88 g/l (normal range 7-16 g/l). An IgG antibody test for *Strongyloides stercoralis* came back negative.

In multiple stool examinations, we were able to find eggs of nematodes – suspected to be of hookworms or *Strongyloides*.

A three-day course of ivermectin was initiated. After the second dose the vomiting stopped and the constipation became better. We performed a second course of ivermectin and additionally mebendazole because we suspected a co-infection with hookworms.

Moreover, we decided to start Vancomycin because we isolated enterococcus faecalis in blood cultures and on the tip of a central venous catheter, we had to place to feed the patient parenterally.

After 24 days the patient was discharged in stable and asymptomatic conditions. By the time of a follow-up visit two weeks later the patient's weight had registered little improvement, with a cachectic 41 kg BMI 16 kg/m<sup>2</sup> but in generally better conditions. Besides a still present hypogammaglobulinemia and b-cell deficiency the laboratory showed

unobtrusively. Especially the albumin and T-cells were normalized. A repeated antibody test for *Strongyloides stercoralis* again showed negative.

Nearly one year later in April 2016 the patient is in a good condition. He has not been hospitalized again. According to his general practitioner he is able to work again and there were no further infections. Even though the patient regained weight he complained about abdominal discomfort. In multiple stool examinations from April 2016 *Strongyloides stercoralis* was found. Because of the natural history of the infection with *Strongyloides stercoralis* we see this as a proof of a Good-Syndrome with a hyper-infection with *Strongyloides stercoralis* after thymomectomy.

### Discussion

The combination of recurrent infection, hypogammaglobulinemia and thymoma is known as the Good Syndrome. It was first described in 1954 by the pediatrician Robert A. Good. It is associated with a significant mortality of 42% as described in a systemic review that looked at 152 cases [1]. The incidence of hypogammaglobulinemia in patients with thymoma is 6% to 11% [2,3]. The syndrome usually presents in the 4<sup>th</sup> or 5<sup>th</sup> decade of life [4] with an equal distribution across the sexes [5]. The spectrum of infections can differ from bacterial, viral or fungal diseases [5]. This case however is a rare case presenting a hyperinfection with *Strongyloides stercoralis* after thymomectomy and furthermore a bacteremia with *Enterococcus faecalis*.

The diagnosis of thymoma can proceed or follow the appearance of immune deficiency or can even be diagnosed simultaneously, which we assume in our case [1]. In contrast to other paraneoplastic syndromes just as Myasthenia gravis, Good syndrome is not resolved by thymectomy. Our patient did show gastrointestinal symptoms before surgery, but the *Strongyloides stercoralis* infection exacerbated severely after thymectomy. We assume that a combination of postoperative stress and the thymectomy itself led to a hyperinfection of *Strongyloides stercoralis* due to an even more compromised immune system. It should be clarified that so far there is only a vague understanding of the pathogenesis. The persistence of hypogammaglobulinemia after thymectomy remains the main obstacle [6].

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Variables	Ref.-range	Unit	29.04.15 pre-Op	15.05.15	20.05.15	16.06.15
Leucocytes	3.90-10.50	mg/nl	5.13	7.60	11.98	7.72
Hemoglobin	13.5-17.0	g/dl	14.3	10.4	11.0	13.6
Thrombocytes	150-170	mg/nl	294	554	531	451
Kreatinin	0.70-1.20	g/dl	0.63	0.55	0.46	0.71
CRP	<5.0	mg/l	0.5	70.1	36.0	1.2
Lymphocytes absolute	1.10-4.50	mg/nl	--	1.04	1.75	3.42
Eosinophils absolute	0.02-0.50	mg/nl	--	0.07	0.11	0.28
Lymphocytes %	22.0-44.0	%	--	13.4	14.6	44.3
Eosinophils %	0.5-5.5	%	--	0.9	0.9	3.6
CD4 helper cells %	38.0-46.0	%	--	46.6	49.2	35.0
CD4 helper cells	700.0-1100.0	mg/μl	--	474.0	860.0	1198.0
CD8 suppressor cells %	31.0-40.0	%	--	28.9	30.1	40.6
CD8 suppressor cells	500.0-900.0	mg/μl	--		527.0	1389.0
CD4/8 Ratio	--	--	--	295.0	1.63	0.86
CD19/B-cells	200.0-400.0	mg/μl	--	130.0	188.0	73.0
CD19 B-cells %	11.0-16.0	%	--	12.8	10.7	2.1
Serum-Elektrophoresis	Ref.-range	Unit	27.05.15	16.06.15		
Albumine	55.8-66.1	%	46.7	57.8		
A1-Globulin	2.9-4.9	%	13.8	7.4		
A2-Globulin	7.1-11.8	%	19.7	13.6		
β-Globulin	8.4-13.1	%	12.2	11.0		
Gamma-Globulin	11.1-18.8	%	7.6	10.2		
Immunoglobulin G	7.00-16.00	g/l	2.88	7.19		
Immunoglobulin A	0.70-4.00	g/l	0.67	1.01		
Immunoglobulin M	0.40-2.30	g/l	0.06	0.33		
IgG4	0.052-1.250	g/l	1.861	0.225		
IgG2	1.150-5.700	g/l	0.465	1.195		

Table 1: Laboratory results.

Especially two different approaches come into consideration. The first one is based on the assumption that there is a direct or indirect crosstalk via cytokines, secreted in the bone marrow between thymus and B cell precursors [7,8]. Another theory keeps antigen-presenting cells (e.g. dendritic cells) as the main target. It could be shown that the amount of DC is decreased in Good Syndrome. So, if antigen presentation to B-cells is reduced, the production of immunoglobulins decreases, which in turn leads to a higher sensitivity to bacterial infections [4].

Even though there was a negative antibody test for *Strongyloides stercoralis* we assume this to be the actual parasite, because of the natural history of the infection. *Strongyloides* is unique because of its ability to survive in the host for decades because of autoinfection mechanisms. Our patient has lived in Germany for 25 years now and has only been back to DRC twice for short stays in hotels.

*Strongyloides* hyperinfections in patients with hypogammaglobulinemia has been described in literature [9,10]. A hyperinfection is defined as an accelerated autoinfection in immunocompromised patients especially in high-dose corticosteroid therapy with an invasion of numerous organs, often leading to infections with bacteria of the gastrointestinal system -like we saw in our patient. Moreover the hyperinfection most often leads to the development of gastrointestinal symptoms in formerly subclinically infected patients. A typical finding in the skin is the larva currens. Hyperinfection shows a mortality rate of up to 90% if untreated. As a sign of severity there is often a lack of eosinophilia [11]. Interestingly this was the case when the patient was admitted to our clinic, in a differential blood count before the surgical intervention an eosinophilia of 16% was observed.

In murine studies, it could be shown that the susceptibility to *Strongyloides* infection is related to the absence of MHC II, which leads

to a decreased Th-2 answer. In fact, MHC II is reduced in patients suffering from thymoma, especially in WHO-type A and AB, which was the case in our patient with a histological WHO-type AB thymoma [12]. This gives us an immunological explanation for the severe nematode infection we saw in our patient.

## Conclusion

Even though Good Syndrome is a very rare disease, it should be considered in patients with unexplained immunodeficiency in context of thymoma. We recommend all patients with thymoma should have B and T cell subsets and immunoglobulin values measured. In addition, a detailed history concerning various infections has to be taken. An abnormal immunoglobulin profile needs further immunological investigation. The treatment of Good Syndrome should be a combination of surgical thymoma resection [13], the treatment of the complications and in some cases an immunoglobulin replacement treatment should be considered.

Because of the complexity of this kind of disease an interdisciplinary approach involving both surgeons and specialists in infectious diseases should be the gold standard.

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