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From a Thymoma Discovered During the Infectious Disease Assessment to the Effects of Thymectomy. Good's Syndrome? A Case Report.

Thierry Sibomana[1]*, Jean Claude Mbonicura[2], Daniel Nduwayo[3], Emile Horugavye[2], Elcy Benita Kaneza[4], Bizoza Yves [2], Francois Nduwimana[2]

1University of Burundi, Kamenge Teaching Hospital (KTH), Pulmonology department.

2University of Burundi, KTH, General Surgery department.

3University of Burundi, KTH, Neurology department.

4University of Burundi, KTH, Internal Medicine department.

Corresponding Author: Thierry Sibomana, University of Burundi, Kamenge Teaching Hospital (KTH), Pulmonology department.

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Abstract

The clinical manifestations of thymoma are highly varied. They can range from an incidental radiological finding to pulmonary signs or those of an upper vena cava syndrome or even signs associated with an autoimmune paraneoplastic syndrome such as the neurological signs found in myasthenia gravis, the autoimmune disease most frequently associated with thymoma. Digestive disorders such as chronic diarrhea and vomiting are rare and can be seen in Good's syndrome associating the presence of thymoma and an acquired immune deficiency. We report a case of a 50-year-old with thymoma with chronic diarrhea and opportunistic infections.

Keywords: *Thymoma, Thymectomy, Opportunistic Infections.*

Introduction:

The thymus is an organ, progressively involuting after puberty, which is involved in the maturation and selection of T cells **[**1**]**. Thymic tumors, including thymoma, thymic carcinoma and thymic neuroendocrine tumors, can be formed from thymic epithelial cells **[**2**]**.

The clinical manifestations of thymoma are highly varied. They can range from an incidental radiological finding to pulmonary signs or those of an upper vena cava syndrome **[**3**]** or even signs associated with an autoimmune paraneoplastic syndrome such as the neurological signs found in myasthenia gravis, the autoimmune disease most frequently associated with thymoma **[**1**]**. Digestive disorders such as chronic diarrhea and vomiting are rare and can be seen in Good's syndrome associating the presence of thymoma and an acquired immune deficiency **[**4**]**.

We report the observation of a patient with thymoma with chronic diarrhea and opportunistic infections. The aim is to show the impact of resection of this thymoma on the evolution of opportunistic infections.

Case Report

A 50-year-old male, the nonsmoker, consulted for chronic motor diarrhea, weight loss of more than 10% over one year, and vomiting for six months. The patient had had a dry, drawn-out cough in the past that he had neglected and that had subsided. Physical examination revealed moderate dehydration and a BMI of 19.59 kg/m². Parasitological examination of the stool and coproculture were normal. Total colonoscopy showed hyperplastic rectal polyposis and a supracentimetric adenomatous polyp of the lower rectum. The colonic biopsy was normal. Thyroid workup was normal. HIV serology was negative. An esophageal fibroscopy had noted esophageal candidiasis. Biology showed anemia of 10.7 g/dl normocytic and normochromic. The sedimentation rate was 52 mm. Biochemistry showed severe renal failure with urea at 7.54 mmol/l and creatinine at 226.18 µmol/l, with a glomerular filtration rate estimated according to Cockroft & Gault at 27.29 ml/min. Venous glucose and blood ionogram were normal. The chest X-ray showed a dense and well-limited opacity in the left pericardiac region. The non-injected chest CT scan showed a left anterior and middle mediastinal, tissue mass measuring 108.7×75.1×153.6 mm with mass effect on the left upper bronchus without evidence of invasion (Image1) suggestive of a thymoma. Protein electrophoresis and an abdominal-pelvic CT scan could not be performed. The patient was rehydrated, put on Fluconazole, intravenous Pantoprazole, Ondansetron and Loperamide without clinical improvement but with normalization of renal function. A surgical procedure was performed by thoracotomy to perform a thymectomy. The patient was positioned in the right lateral decubitus position and the approach taken was a left anterolateral thoracotomy. After dissection of the chest wall and exposure of the mediastinum with a Finochietto retractor (Image 2 and 3), we found a multilobulated anterior mediastinal mass. Dissection of the mass was performed (Image4

and 5) and removal of the mass (Image6) was completed by removal of the surrounding fatty tissue and warm saline lavage followed by parietal closure.

Pathological examination of the surgical specimen concluded an encapsulated thymoma of type AB (according to the World Health Organization 2016 classification) with a weight of 616g and measuring 17 x 10 x 5cm. The presence of thymoma and a clinical picture of immunosuppression made us think of Good's syndrome.

The patient could not receive immunoglobulin injection, not available locally. The evolution was marked by persistent episodes of diarrhea and vomiting.

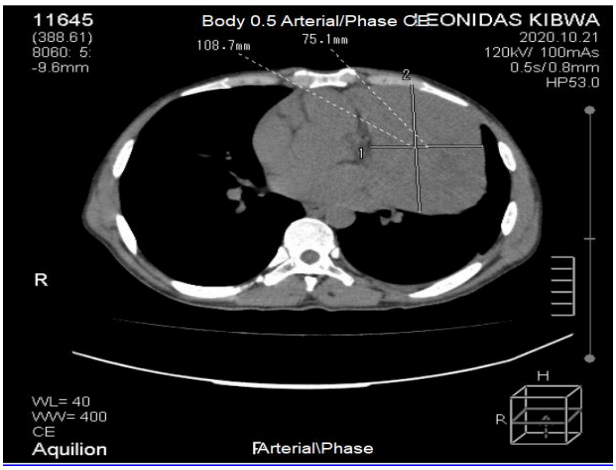

Image 1: Non-injected chest CT scan. Anterior and middle left mediastinal mass, tissue measuring 108.7×75.1×153.6mm with mass effect on the left upper bronchus with no evidence of invasion

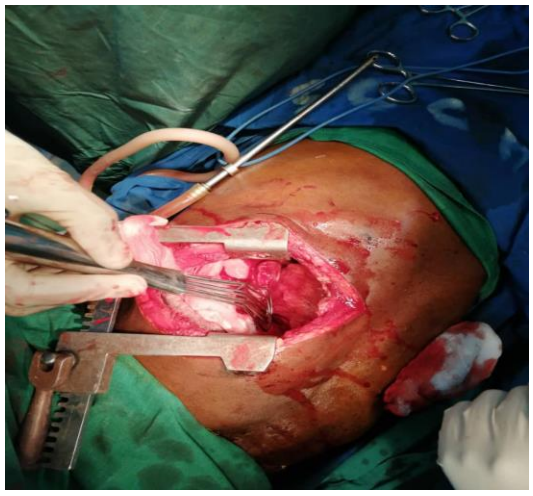

Image2: Intrathoracic mass in our patient in the right lateral decubitus position

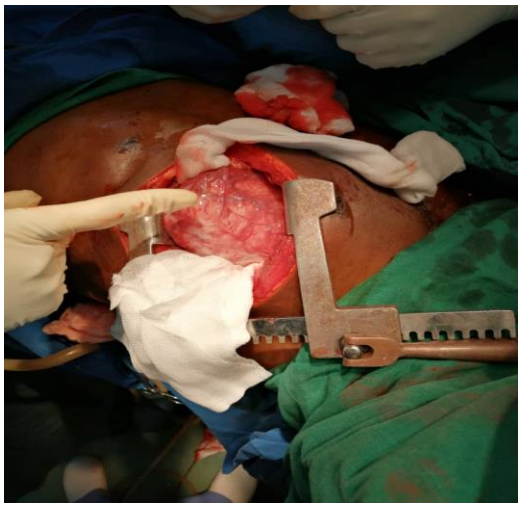

Image3: Intrathoracic mass well exposed for excision

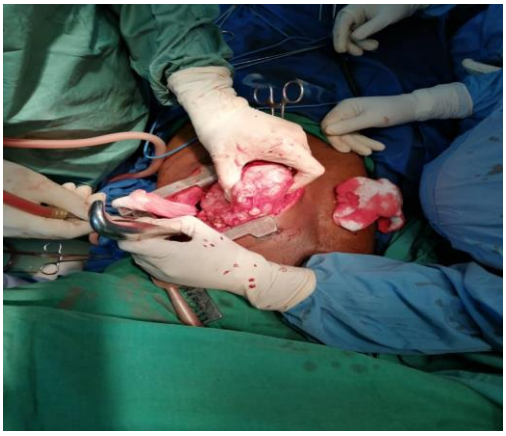

Image 4: Tumor removal in progress

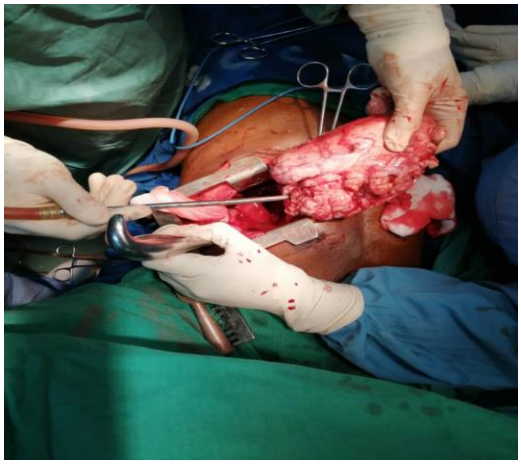

Image 5: Release of the last tumor attachments

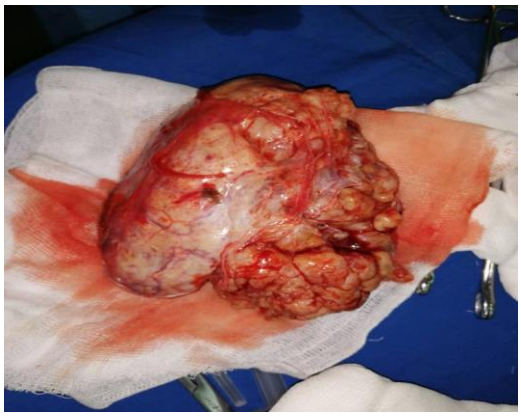

Image 6: Macroscopic appearance after resection of an anterior mediastinal mass: Tumor mass weighing 616 g and measuring 17 x 10 x 5cm, multilobulated**.**

Discussion

Despite their rarity, thymomas represent 50% of anterior mediastinal tumors **[**5**]**. They can occur at any age but affect equally males and females between the ages of 40 - 60 years **[**2**]**.

The circumstances of discovery are varied. Asymptomatic in 30-50% of cases, thymoma is often discovered incidentally during a radiological examination or even during thoracic surgery **[**3,6**]**. The search for pulmonary infectious disease associated with chronic diarrhea and esophageal candidiasis led to the discovery of thymoma in our case.

The thoracic CT scan with contrast medium is the standard diagnostic examination for thymoma **[**3**]**, a tool for preoperative staging of thymoma **[**7**]** conditioning its treatment, for monitoring after its resection **[**6**]** but also informing us about its prognosis **[**7**]**. An anterior mediastinal mass could already be evoked by the demonstration of a dense and well-limited opacity at the left pericardiac level on the thoracic radiograph. As for the scenographic findings, they allowed us to determine the delimitation of the mass about the neighboring structures and the absence of signs of invasion and therefore the possibility of a total resection.

The encapsulated thymoma type AB (according to the World Health Organization 2016 classification) found on anatomopathological examination, in our patient, is the most frequent, depending on the series, or the second most frequent thymoma in the literature **[**8**]**. The histological classification associated with the staging of the tumor (Masaoka-Koga classification and TNM classification) **[**1**]** has a prognostic value. A, AB and B1 tumors have no (stage I and II) to low (stage III) malignant potential **[**8**]**.

In general, because of the malignant potential of all thymomas, their total resection is preferable **[**6**]** and is associated with an excellent prognosis **[**9**]** in the short and long term **[**3**]**. However, a multidisciplinary

consultation is always recommended [10]. For advanced thymoma (Masaoka-Koga stage III and IVA), induction chemotherapy followed by en bloc resection, when a sufficient tumor response is obtained, is performed in a curative approach [10].

The picture of an immune deficiency manifested by chronic diarrhea and esophageal candidiasis is by far the one that could have directed us to thymoma. However, an entity, Good's syndrome [4], has been described in the literature, associating thymoma, acquired hypogammaglobulinemia of the different immunoglobulin classes and, in almost all cases, B lymphocytopenia. The latter may explain the susceptibility to infections [11] and even opportunistic infections similar to those observed in AIDS, in particular cytomegalovirus infections, candidiasis and disseminated tuberculosis [11].

The treatment of choice for thymoma is surgical resection, which is associated with a mortality rate of 2% and a morbidity rate of 20%. In our patient, a left anterolateral thoracotomy thymectomy was performed. This was an en bloc thymectomy extended to the peri-thymic and anterior mediastinal fat respecting the phrenic nerve.

Phrenic nerve preservation is particularly crucial in patients with myasthenia gravis, but increases the risk of local recurrence, without altering patient survival [12].

Sternotomy is another preferred approach by several authors. However, minimally invasive techniques such as video-assisted thoracoscopy and robot-assisted thoracic surgery (technical platform not allowing it locally) are more and more used. In particular, robot-assisted thoracic surgery offers very good exposure and precision in the resection of thymoma [13].

However, Y. Jamilloux et al, in their review of autoimmune diseases associated with thymoma, found that surgical resection did not alter the course of autoimmune manifestations [1]. This could explain the persistence of digestive disorders in our patients after thymectomy. Furthermore, given the significant infectious morbidity in patients with thymoma, immunoglobulin replacement therapy is recommended in those with thymoma with hypogammaglobulinemia and those receiving immunosuppressive therapy, even in the absence of prior infection [11].

Conclusion

Thymoma is one of the pathologies to be considered during the assessment of digestive disorders such as chronic diarrhea and vomiting associated with respiratory complaints. Surgical resection may not alter the course of already established autoimmune manifestations and opportunistic infections.

References

1. Jamilloux Y, Frih H, Bernard C, Broussolle C, Petiot P, Girard N, et al. Thymomes et maladies auto-immunes. La Rev médecine interne [Internet]. 2018;39(1):17–26. Available from: http://dx.doi.org/10.1016/j.revmed.2017.03.003

2. Engels EA. "Epidemiology of thymoma and associated malignancies". J Thorac Oncol. 2010;5(10 Suppl 4):S260-5.

3. Jungraithmayr W, Schneiter D, Soltermann A, Frauenfelder T, Weder W. Le thymome. "Forum Médical Suisse" – Swiss Med Forum. 2016;16(45):961–8.

4. Kelesidis T, Yang O. Good's syndrome remains a mystery after 55 years: A systematic review of the scientific evidence. Clin Immunol [Internet]. 2010;135(3):347–63. Available from: http://dx.doi.org/10.1016/j.clim.2010.01.006

5. Tian D. "Tumor location may affect the clinicopathological features and prognosis of thymomas". 2019;10:2096–105.

6.Detterbeck FC, Zeeshan A. "Thymoma : current diagnosis and treatment". Chin Med J. 2013;126(11):2186–91.

7. Qu Y juan, Liu G bing, Shi H shui, Liao M yan, Yang G fang, Tian Z xiong. Preoperative CT Findings of Thymoma are Correlated with Postoperative Masaoka Clinical Stage. Acad Radiol [Internet]. 2013;20(1):66–72. Available from: http://dx.doi.org/10.1016/j.acra.2012.08.002

8. Parrens M. Pathologie du médiastin . Cas No 3 . Thymome de type AB. Ann Pathol [Internet]. 2015;35(3):224–9. Available from: http://dx.doi.org/10.1016/j.annpat.2015.03.007

9. Safieddine N, Liu G, Cuningham K, Ming T, Hwang D, Brade A, et al. Prognostic factors for cure, recurrence and long-term survival after surgical resection of thymoma. J Thorac Oncol [Internet]. 2014;9(7):1018–22. Available from: http://dx.doi.org/10.1097/JTO.0000000000000215

10. RYTHMIC R tumeurs Thym et C. "Référentiel de prise en charge des tumeurs thymiques". 2020;

11. Malphettes M, Gérard L, Galicier L, Boutboul D, Asli B, Szalat R, et al. "Good Syndrome: An Adult-Onset Immunodeficiency Remarkable for Its High Incidence of Invasive Infections and Autoimmune Complications". Clin Infect Dis. 2015;61(2):e13–9.

12. Hamdi S, Mercier O, Fadel E, Mussot S, Fabre D, Ghigna MR, et al. "Is sacrifying the phrenic nerve during thymoma resection worthwhile?" Eur J Cardiothorac Surg 2014; 45:e151-5.

13.Schneiter D, Tomaszek S, Kestenholz P, Hillinger S, Opitz I, Inci I, Weder W. “Minimally invasive resection of thymomas with the da Vinci(R) Surgical System”. Eur J Cardiothorac Surg. 2013;43:288–92.