

pulmonary aneurysm is well described in the literature and is usually successful in treating RA.^{1,5} Possible complications of treatment are bleeding recurrence, inadvertent embolization of a spinal artery leading to ischemic myelopathy, and aneurysmal rupture during diagnostic catheter angiography,^{1,5} with a high risk of mortality. Surgical excision is recommended only when expert radiological intervention is not available or in the presence of considerable destructive process in the lung.⁵

In conclusion, Rasmussen aneurysm should be included in the differential diagnosis of hemoptysis in patients with TB. Enhanced CT examination plays an important role in the evaluation of these patients.

References

1. Peghini Gavilanes E, López Yepes LA, Peñalver Paolini CL, Morales Ruiz R. Rasmussen's pseudoaneurysm in a patient with a history of pulmonary tuberculosis. *Arch Bronconeumol.* 2015;51:96–7.

2. Bruzzi JF, Rémy-Jardin M, Delhay D, Teisseire A, Khalil C, Rémy J. Multi-detector row CT of hemoptysis. *Radiographics.* 2006;26:3–22.
3. Keeling AN, Costello R, Lee MJ. Rasmussen's aneurysm: a forgotten entity. *Cardiovasc Interv Radiol.* 2008;31:196–200.
4. Castañer E, Gallardo X, Rimola J, Pallardó Y, Mata JM, Perendreu J, et al. Congenital and acquired pulmonary artery anomalies in the adult: radiologic overview. *Radiographics.* 2006;26:349–71.
5. Wang W, Gao L, Wang X. Rasmussen's aneurysm with aspergilloma in old, healed pulmonary tuberculosis. *Clin Imaging.* 2013;37:580–2.

Rafael Domingos-Grando,^a Gláucia Zanetti,^b Edson Marchiori^{b,*}

^a Hospital Nossa Senhora Conceição, Puerto Alegre, Brazil

^b Federal University of Rio de Janeiro, Rio de Janeiro, Brazil

* Corresponding author.

E-mail address: edmarchiori@gmail.com (E. Marchiori).

Combined Alpha-1-Antitrypsin Deficiency and Mannose-Binding Lectin Deficiency[☆]



Déficit combinado de alfa-1-antitripsina y déficit de lectina fijadora de manosa

To the Editor,

Alpha-1-antitrypsin deficiency (AATD) is a hereditary disease. Clinical manifestations include pulmonary emphysema, hepatic cirrhosis, and more rarely, ANCA-positive panniculitis or vasculitis.¹ Few instances of its association with other genetic deficits have been reported in the literature.

We report the case of a 36-year-old man with no toxic habits and a history of atrial fibrillation, treated with oral anticoagulants, referred to the pulmonology department for genetic counselling after the detection of a family member as an AATD index case (father with SZ genotype). Genetic studies were performed, and the patient was diagnosed with type SZ AATD, with serum protein levels of 46.4 mg/dl. Spirometry and high-resolution computed tomography were normal. In subsequent follow-up visits, he began to present repeated bronchial infections, so a full study of primary immunodeficiencies was performed. In this study, very low levels of mannose-binding lectin (MBL) were detected on ELISA (normal range >1000 ng/ml, deficit considered <500 ng/ml and severe deficit <100 ng/ml): the initial result was 7.54 ng/ml, and a subsequent test found 72.807 ng/ml. Other primary immunodeficiency results, including immunoglobulin serum levels, were normal (IgG 1186 mg/dl; IgA 362.80 mg/dl; IgM 75.60 mg/dl).

Alpha-1-antitrypsin (AAT) is a glycoprotein¹ synthesized mainly in the liver. Its principal function is to inhibit neutrophil elastase and other proteases, and its anti-inflammatory, anti-microbial and immunomodulatory properties have been characterized in recent years. AAT deficiency is caused by a mutation on the long arm of chromosome 14. Lectin is a protein synthesized in the liver which is found in different types of biological fluids, and is considered by some authors as an acute-phase reactant.² MBL is involved in the activation of the lectin complement system, which has been established as the third activation pathway.³ MBL deficiency is caused by a mutation on the long arm of chromosome 10. Low serum

levels of MBL are associated with an increased risk of various infections in children. In adults, this deficiency may be associated with concomitant diseases or immunodeficiencies.⁴ MBL deficiency has been associated with predisposition to a large number of diseases (autoimmune diseases, in particular), but it generally has more clinical significance when it coexists with another immunodeficiency state.⁵ Curiously, the association of MBL deficiency and AATD in the same patient has not been described previously.

The clinical presentations of AATD vary widely, so disease registries and studies are needed to better determine its natural history. Taking into account the prevalence of both deficiencies in the general population, its association in our patient may be a random event. Nevertheless, these immunodeficiencies may need to be ruled out in AATD patients, since they could modify the clinical manifestation and management of the disease, as occurs with other immunodeficiency states.

References

1. American Thoracic Society; European Respiratory Society. American Thoracic Society/European Respiratory Society statement: standards for the diagnosis and management of individuals with alpha-1 antitrypsin deficiency. *Am J Respir Crit Care Med.* 2003;168:818–900.
2. Holmskov U, Thiel S, Jensenius JC. Collections and ficolins: humoral lectins of the innate immune defense. *Annu Rev Immunol.* 2003;21:547–78.
3. Takahashi M, Iwaki D, Kanno K, Ishida Y, Xiong J, Matsushita M, et al. Mannose-binding lectin (MBL)-associated serine protease (MASP)-1 contributes to activation of the lectin complement pathway. *J Immunol.* 2008;180:6132–8.
4. Summerfield JA, Ryder S, Sumiya M, Thursz M, Gorchein A, Monteil MA, et al. Mannose binding protein gene mutations associated with unusual and severe infections in adults. *Lancet.* 1995;345:886–9.
5. Chalmers JD, McHugh BJ, Doherty C, Smith MP, Govan JR, Kilpatrick DC, et al. Mannose-binding lectin deficiency and disease severity in non-cystic fibrosis bronchiectasis: a prospective study. *Lancet Respir Med.* 2013;1:224–32.

Cecilia López-Ramírez,^{a,*} José Luis López-Campos^{a,b}

^a Unidad Médico-Quirúrgica de Enfermedades Respiratorias, Hospital Universitario Virgen del Rocío, Instituto de Biomedicina de Sevilla, Sevilla, Spain

^b CIBER de Enfermedades Respiratorias (CIBERES), Instituto de Salud Carlos III, Madrid, Spain

* Corresponding author.

E-mail address: ceclopram@gmail.com (C. López-Ramírez).

[☆] Please cite this article as: López-Ramírez C, López-Campos JL. Déficit combinado de alfa-1-antitripsina y déficit de lectina fijadora de manosa. *Arch Bronconeumol.* 2016;52:174.