Life-threatening Misdiagnosis of Bulbar Type Onset Myasthenia Gravis as a Subacute Thyroiditis

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Myasthenia gravis (MG) is an autoimmune disease that can mimic a variety of symptoms leading to a delay in diagnosis and treatment. We report the case of 48 year-old woman, in whom initial presentation of MG with predominance of bulbar symptoms was mistaken for thyroid disease complications. Subsequently, correct diagnosis and optimal management resulted on significant improvement of her functional state. We discuss the importance of considering MG as one of the potential differential diagnoses among cases of new onset or recurrent unexplained bulbar symptoms. [P R Health Sci J 2019;38:120-121]

Key words: Myasthenia gravis, Thyroid carcinoma, Bulbar symptoms

yasthenia gravis (MG) is an autoimmune disease that affect the neuromuscular junction (NMJ) in which most antibodies are directed against the acetylcholinesterase receptor (AChR). With continued activation of the nerve, the safety margin for neuromuscular transmission is lowered at an increasing number of NMJs and many muscle fibers fails to activate, causing variable degrees of weakness and easily fatigability of voluntary skeletal muscles. Weakness appears to worsen with sustained exercise of extraocular muscles (diplopia) bulbar muscles (dysarthria, dysphagia), and limb muscle fatigability. MG is rarely reported to be associated with thyroid tumors, although MG is sometimes accompanied by neoplasm, especially thymoma (1,2). Maruyama et al., showed that the prevalence of thyroid carcinoma in MG may be higher than in general population and frequently coexists with chronic thyroiditis (1). Thyroiditis is probably secondary to the presence of the tumor, potentially owing to an autoimmune response to the antigen exposed by the tumor (2,3). However, the 25 % of patient with MG produces a variety of autoantibodies, including antithyroid antibodies that may be related to thyroiditis, which may mimic a variety of thyroid disease's symptoms leading to a delay in diagnosis and treatment (2). Moreover, thyroid disease may preceded the neuromuscular disorder in MG patients, presenting initially with thyrotoxicosis with subsequent development of hypothyroidism (3). Hence, when the two diseases exist simultaneously (i.e., thyroid disease in MG patient) either one may easily be overlooked.

Case Report

A 48 years old woman with history of fibromyalgia and multinodular goiter presented to emergency department (ED) of our University Hospital in February 2017 with complaint of

progressive dysphagia initially to solid and then to liquid and solids, odynophagia, anorexia, hoarseness, and sudden thyroid swelling that worsen nine days prior to admission that forced her to seek medical attention. Patient also reports episodes of tremors, palpitations, sweating, fatigue, dyspnea and anxiety that worsen during the last week prior to admission. The muscles of jaw closure were involved and produced weakness with prolonged meal time (fatigable chewing). All symptoms started after fine needle aspiration (FNA) biopsy of two nodules (> 1cm) performed one week prior to admission, which resulted in poor oral intake and dehydration. FNA biopsy was positive for follicular thyroid carcinoma. On physical examination, patient was found with enlarged thyroid with asymmetric multinodular goiter and very tender upon palpation and no ocular symptoms (ptosis and/or diplopia), nasal speech or head drop. Neurological exam is remarkable for no facial asymmetry, and limb preserved strength with no fatigability with sustained exercise. EKG remarkable for sinus tachycardia. Neck CT scan revealed heterogenous enlarged thyroid gland with multi-septate left lobe complex lesion and right lobe mass. Laryngoscopy showed no pathology and intact vocal cords. The impression diagnosed of subacute thyroiditis causing tracheoesophageal compression syndrome was established on the basis of symptoms presented and in the light of her previous history, as well as evidenced by her thyroid function

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test. Laboratories results showed normal low border TSH, increase Thyroxine (T4) and Triiodothyronine (T3) uptake, elevated ESR and CPR, detected levels of thyroglobulin and undetected thyroglobulin antibodies. The patient received treatment with intravenous steroids showing markedly improvement of bulbar symptoms (dysphagia, and hoarseness). At the moment of discharge, she was clinically euthyroid with normal values for serum concentration of T4, T3 and TSH. Patient continue ambulatory steroid tapering treatment, but after one month, ocular symptoms and generalized proximal muscle weakness was added to her symptoms. In April 2017, patient was urgently admitted to our University Hospital because progressive dysphagia, hoarseness, ptosis, diplopia, weakness of her jaw and proximal limbs, and respiratory failure requiring mechanical ventilation intubation. The diagnosis of MG was suspected on basis of neurological symptoms and subsequently established by positive antiacetylcholine receptor antibody. Chest CT scan revealed thyroid gland with multi-septate left lobe complex lesion and well-defined lesion at thyroid isthmus and no evidence of mediastinal mass. Her ocular and bulbar symptoms, as well as her limb weakness improved with intravenous immunoglobulin treatment, pyridostigmine and corticosteroids therapy. Patient was extubated in a few days and discharged with significant improvement in her functional state. At 4 months of follow-up, the patient is doing well on pyridostigmine and corticosteroids therapy.

Discussion

Our patient, with history of multinodular goiter and recently post thyroid biopsy with suspicious follicular nodules, lack of obvious ocular findings, and presenting with prominent bulbar symptoms led to incorrect interpretation of the findings. Furthermore, the initial presentation of thyrotoxicosis signs, as well as imaging and laboratory findings deviated the attention toward to interpret observations as thyroiditis [induced by trauma (FNAB)] with associated swelling of goiter, causing obstructive and bulbar symptoms. Hence, this case illustrates the diagnostic challenge posed by MG and the importance of bearing in mind this diagnosis in the case of patients presenting with bulbar symptoms. About 15% of the patients present with only focal bulbar weakness as initial presentation, especially in the late age-onset of MG (4). MG and hyperthyroidism are strongly associated and often develop simultaneously (2). Moreover, chronic thyroiditis has been found in up to 19% among MG patients with coexistent adenomatous goiter (2). Therefore, such atypical presentation of bulbar symptoms, in the absence of obvious ocular involvement, does not exclude MG. Literature revealed that initially misdiagnosed MG cases had atypical presentation in the absence of ocular symptoms and limb girdle myasthenia, leading to a delay in the ultimate diagnosis and prompting consideration of other diagnostics (5).

In our patient, there was a delay of several months in reaching diagnosis. This delay was compounded by the following issues: 1) presence of bulbar palsy at the start of disease, in the absence of any ocular symptoms, 2) coexistence of bulbar and thyrotoxicosis symptoms, and 3) history of multinodular adenomatous goiter and recent thyroid biopsy that mislead attention to thyroid disease complications, causing swelling of thyroid gland and obstructive symptoms. However, reappearing of the bulbar symptoms and added ocular and proximal muscle weakness findings after completing steroid therapy led to suspicious MG and subsequent diagnosis.

Conclusion

In focal bulbar weakness, a high index of suspicious is required to MG. The presented case shows that it is important to consider MG in patients with history of thyroid disease and presenting with progressive bulbar muscle weakness, even with thyrotoxicosis signs.

Resumen

La variedad de síntomas que se presentan en los pacientes con la enfermedad autoinmune de Miastenia Gravis (MG) pueden desviar su diagnóstico correcto, ya que pueden disfrazar el padecimiento con otros diagnósticos. En este documento presentamos los hechos del caso de una mujer de 48 años, que comenzó mostrando síntomas bulbares que fueron confundidos por un agravamiento de una enfermedad tiroidea. Subsecuentemente, un diagnóstico correcto y cuidado óptimo devengaron en una mejoría significativa en su estado funcional. Discutimos aquí, la importancia de considerar MG cómo un diagnóstico diferencial potencial en los nuevos casos que se presentan a sala y aquellos recurrentes en que se muestren inexplicables síntomas bulbares.

References

- Maruyama E., Minami N., Niino M., Fujiki N., Doi S., Watanabe M., Shima K., Kikuchi S., Sasaki H. Impact of screening with thyroid ultasonograpghy in myasthenia gravis patients. Acta Neurol Scand 2011;125:398-402.
- Donaldson J. O., Margaret L., Grunnet L., Thompson H. G. Concurrence of myasthenia gravis, thymoma, and thyroid carcinoma Arch Neurol 1983;40:122-124.
- 3. Aarli J. A., Thunold S., Heimann P. Acta Neurol Scand 1978;58:121-127.
- Oosterhuis H.J., The natural course of myasthenia gravis: A long term follow up study. J Neurol Neurosurg Psychiatry 1989;52:1121-1127.
- Wheeler S.D. Misdiagnosis of myasthenia gravis J Natl Med Assoc 1987;79:425-334.