Can't Open My Eyes and Have Trouble Breathing: An Atypical Presentation of Myasthenia Gravis Leading to Interstitial Lung Disease

Kaur Harjeet, MD; Ubaldo R. Madera Sánchez, MD; Juan Flores, MD.
Hospital Universitario Ramon Ruiz Arnau / Universidad Central del Caribe, Internal Medicine Residency Program Puerto Rico.

Introduction

Myasthenia gravis is an autoimmune disease in which antibodies directed against post-synaptic nicotinic acetylcholine receptors attack the myoneural junction and damage the post-synaptic membrane. We are presenting a 37 years-old male that presented Myasthenia Gravis with atypical presentation of dyspnea in uncommon gender with no proximal muscle weakness and that developed interstitial lung disease.

Clinical Case

A 37 years-old male with no past medical history reported having a two-month history of fluctuating dyspnea on exertion, which progressed to dyspnea at rest. One day during work, the patient had severe shortness of breath associated with blurry vision and could not open his eyes. During his visit to his primary doctor his clinical exam was unremarkable except bilateral ptosis. His medical doctor ordered routine laboratories, Complete blood count, Comprehensive Metabolic Profile, and Thyroid profile which were normal. Chest x-ray and Head MRI were unremarkable. The patient continued with dyspnea and could not open his eyes for weeks. The patient was then referred to a neuro-ophthalmologist and Neurology. Neuro-ophthalmologist performed an Ice pack test, and the neurologist ordered serological studies.

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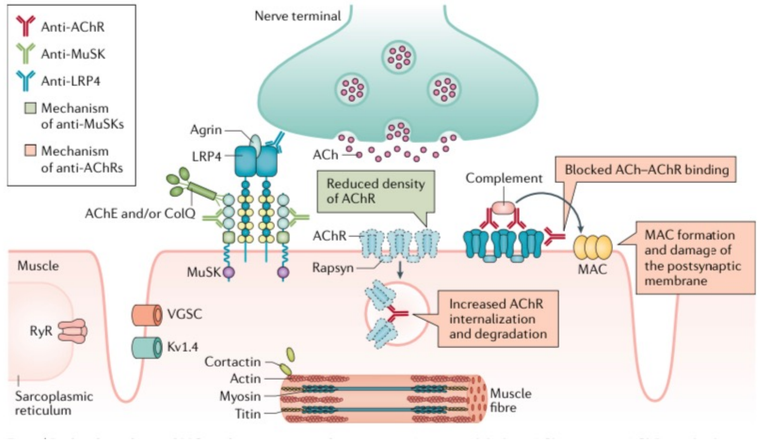
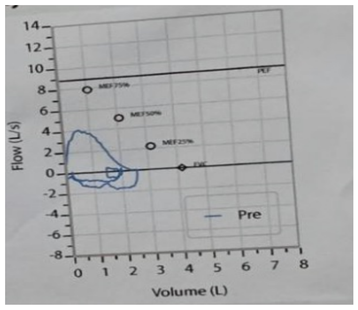
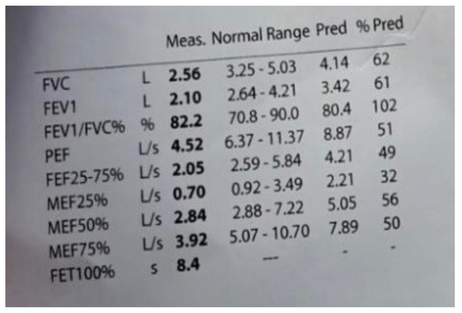
Pathophysiology of Myasthenia Gravis



Pulmonary Function Test





		Meas.	Normal Range	Pred	% Pred
FVC	L	2.56	3.25 - 5.03	4.14	62
FEV1	L	2.10	2.64 - 4.21	3.42	61
FEV1/FVC%	%	82.2	70.8 - 90.0	80.4	102
PEF	L/s	4.52	6.37 - 11.37	8.87	51
FEF25-75%	L/s	2.05	2.59 - 5.84	4.21	49
MEF25%	L/s	0.70	0.92 - 3.49	2.21	32
MEF50%	L/s	2.84	2.88 - 7.22	5.05	56
MEF75%	L/s	3.92	5.07 - 10.70	7.89	50
FET100%	s	8.4	---	-	-

Interpretation : The subject has a FEV1/FVC% ratio of 82.2% which is within normal limits while a FVC value of 2.56 L is below the normal. The presence of Moderate Restriction is confirmed by Total Lung Capacity value of 4.63 L which is below the normal range and a FEV1 value of 2.10 within the range of 60%-90% predicted.

Analysis and Results

Non-invasive Ice Pack Test: Positive
Routine Laboratory work: CBC,CMP,HBAIc,T4 Free TSH: Negative
Serological studies: Serologic tests for Acetylcholine receptor antibodies 0.30 nmol/L (normal <0.40 nmol/L), blocking antibody was 15 (normal <26%), and Striated muscle ab screen was negative
Imaging: Head CT/Chest CT/MRI of Brain: Unremarkable
Pulmonary Function testing: Pulmonary Function Tests were abnormal with decreased FVC 2.56 L, TLC 4.63L, findings consistent with moderately restricted defect and interstitial lung disease.
The patient was prescribed Pyridostigmine and steroids, which led to significant blurry vision and ptosis relief. Interestingly the patient did not have sufficient therapeutic response on first-line agents and required immunoglobulins and corticosteroids treatment. After this treatment, the patient had a considerable improvement in his symptoms.

Conclusion

This case highlights the importance of a high clinical index of suspicion for Myasthenia Gravis despite having seronegative classic markers in patients with unexplained dyspnea, blurry vision, and ptosis, even when there is no classical fluctuating proximal muscle weakness. Atypical cases of Myasthenia Gravis may lead to delayed diagnosis and effects the quality of life or lead to life-threatening conditions in young patients.

References

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