

Neurology

Clinical Feature:

Medical History: A 60-year-old man

Well-controlled hypertension, hyperlipidemia, and type 2 diabetes mellitus

History of Present Illness: 2 month history of progressive, symmetric muscle ache in the proximal arms and legs, difficulty rising from deep chairs, and difficulty climbing stairs. He reports no skin rashes, joint pain, or shortness of breath

Medication List: Atorvastatin (40 mg daily) for the past 3 years, lisinopril, and metformin

Neurological Examination: Normal cranial nerves. Sensation, coordination, and deep tendon reflexes are completely normal. Symmetric, moderate weakness of proximal upper and lower extremities, and neck flexion (graded 4/5 on the MRC scale). Normal distal limb strength. He requires the use of his arms to push himself up from a chair. No skin changes are noted

Initial Laboratory Results: CK: 8,500 U/L (normal: <192 U/L). AST: 380 U/L (normal: <35 U/L)

ALT: 410 U/L (normal: <45 U/L). GGT: Completely normal

CBC and TSH are normal

STEP 1 —

Question: Based on the clinical presentation, exam findings, and initial laboratory results, what is the most appropriate initial diagnostic interpretation and clinical action?

(بدون محدودیت در انتخاب گزینه‌ها - انتخاب گزینه غلط، نمره منفی دارد)

مدت آزمون: 20	آزمون: 18_بیماریهای مغز و اعصاب_PMP
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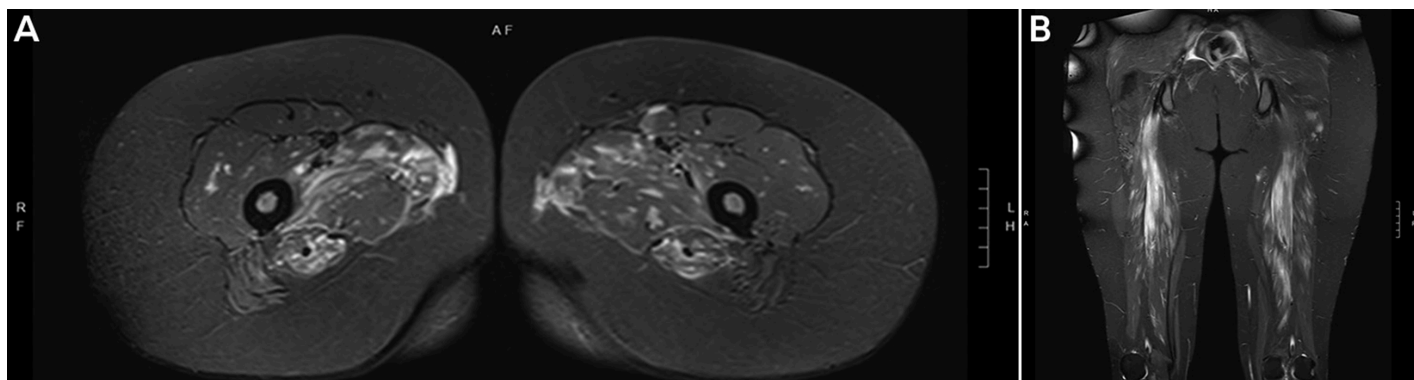
مرگ	عدم انتخاب	نمره انتخاب	بازخورد	گزینه
	0.00	3.00	Correct. Discontinuing the statin is the critical first step.	Discontinue atorvastatin immediately
	0.00	0.00	Incorrect	Initiate high-dose oral Prednisone (1mg/kg, i.e., 80mg daily) immediately to treat suspected polymyositis.
	0.00	2.00-	Incorrect. The patient's normal (GGT confirms that the highly elevated AST and ALT are leaked from damaged skeletal muscle, not the liver	Refer the patient for an emergent liver biopsy to evaluate for acute atorvastatin-induced liver injury.

	0.00	1.00	Correct. Normal	Serum urea and creatinine measurement
	0.00	1.00-	Incorrect. EMG cannot differentiate between specific inflammatory myopathy subtypes and may cause local muscle injury that compromises subsequent biopsy	Order an urgent needle (EMG) of the weak quadriceps muscle to differentiate between inflammatory myopathy subtypes
	0.00	2.00-	Dangerous. Continuing statin therapy in a patient with active, severe myotoxicity with a CK of 8,500 U/L carries an extremely high risk of precipitating acute rhabdomyolysis and secondary acute kidney injury.	Recommend no changes to his medications and refer him to physical therapy for aerobic conditioning.

STEP 2—

Case Evolution: The patient returns to your clinic 2 weeks after stopping atorvastatin. He reports that his proximal weakness has significantly worsened, and he now requires a wheelchair to travel long distances. A repeat laboratory panel is performed, demonstrating a serum CK of 12,800 U/L.

An axial STIR MRI of his thighs is performed, demonstrating diffuse, bilateral symmetric hyperintensity (edema) in the quadriceps and adductor compartments, as illustrated below:



Question: Given his clinical progression and rising CK despite statin cessation, what is the most high-yield diagnostic test to order

(بدون محدودیت در انتخاب گزینه‌ها - انتخاب گزینه غلط، نمره منفی دارد)

مرگ	عدم انتخاب	نمره انتخاب	بازخورد	گزینه
	0.00	3.00	Correct: 315 U (normal: <20 U) intensity 4+	Order serum anti-HMGCR antibody
	0.00	1.00-	Not performed	Order serum anti-Mi-2 antibodies
	0.00	1.00-	Not performed	Order serum anti-Jo-1 antibodies
	0.00	1.00-	Not performed	Order serum anti-MDA-5

0.00	1.00	Correct: negative	Order serum anti-SRP
0.00	1.00-	Not performed	Schedule a biopsy of his severely weak right iliopsoas muscle (MRC grade 3/5) to maximize diagnostic histopathological yield.

STEP 3 —

Case Evolution: The serum anti-HMGCR antibody panel returns **highly positive at 315 U (normal: <20 U)**. An open muscle biopsy of his mildly weak left biceps brachii confirms the diagnosis of anti-HMGCR immune-mediated necrotizing myopathy (IMNM).

The patient is highly anxious to start therapy to regain his mobility but notes that his type 2 diabetes has been extremely difficult to manage, with a recent admission for diabetic ketoacidosis (DKA) and a baseline Hemoglobin A1c of 10.5%.

Question: What is the most appropriate first-line therapeutic regimen for this patient to achieve clinical remission while minimizing systemic, life-threatening complications?

(بدون محدودیت در انتخاب گزینه‌ها - انتخاب گزینه غلط، نمره منفی دارد)

مرگ	عدم انتخاب	نمره انتخاب	بازخورد	گزینه
	0.00	3.00	Correct.	Initiate Intravenous Immunoglobulin (IVIg) 2 g/kg monthly as monotherapy, with serial monitoring of clinical strength and serum CK levels
	0.00	0.00	In a patient with poorly controlled type 2 diabetes and a history of DKA, high-dose corticosteroids carry an extremely high risk of precipitating life-threatening diabetic ketoacidosis or hyperglycemic hyperosmolar state.	Initiate high-dose intravenous methylprednisolone (1 g daily) for 5 days, followed by oral prednisone 80 mg daily, while withholding all other immunotherapies.
	0.00	3.00-	Dangerous & Not performed.	Re-prescribe atorvastatin at a lower dose (10 mg daily) to induce immunological tolerance
	0.00	2.00-	Incorrect. Not performed	Prescribe oral Pyridostigmine 60 mg three times daily to improve neuromuscular transmission at the motor endplates.
	0.00	0.00	RTX is a good second-line or add-on therapy in biopsy-confirmed anti-HMGCR IMNM	Start Rituximab 1000mg two doses, 2 weeks apart

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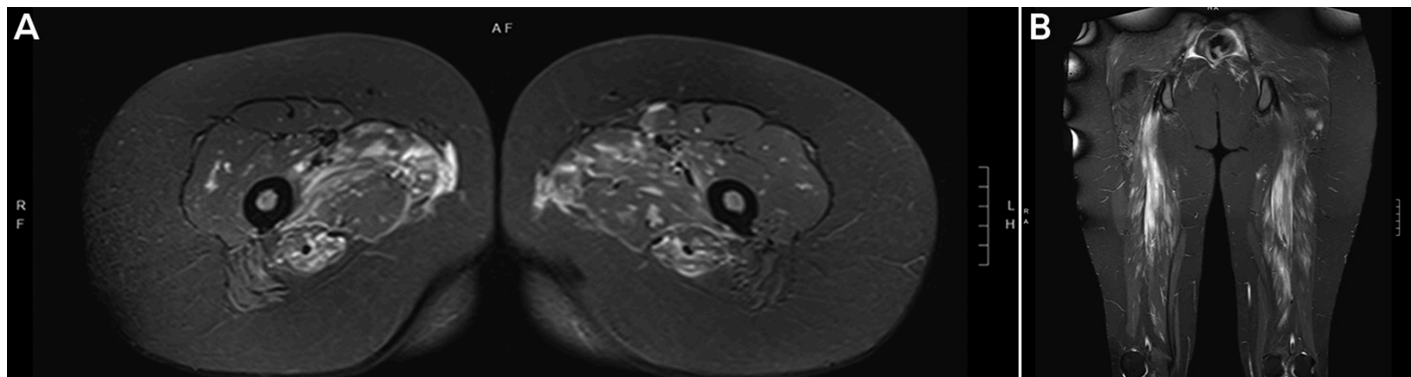
گزینه	بازخورد	نمره انتخاب	عدم انتخاب	مرگ
Discontinue atorvastatin immediately	Correct. Discontinuing the statin is the critical first step.	3.00	0.00	
Initiate high-dose oral Prednisone (1mg/kg, i.e., 80mg daily) immediately to treat suspected polymyositis.	Incorrect	0.00	0.00	
Refer the patient for an emergent liver biopsy to evaluate for acute atorvastatin-induced liver injury.	Incorrect. The patient's normal (GGT confirms that the highly elevated AST and ALT are leaked from damaged skeletal muscle, not the liver	2.00-	0.00	
Serum urea and creatinine measurement	Correct. Normal	1.00	0.00	

	0.00	1.00-	Incorrect. EMG cannot differentiate between specific inflammatory myopathy subtypes and may cause local muscle injury that compromises subsequent biopsy	Order an urgent needle (EMG) of the weak quadriceps muscle to differentiate between inflammatory myopathy subtypes
	0.00	2.00-	Dangerous. Continuing statin therapy in a patient with active, severe myotoxicity with a CK of 8,500 U/L carries an extremely high risk of precipitating acute rhabdomyolysis and secondary acute kidney injury.	Recommend no changes to his medications and refer him to physical therapy for aerobic conditioning.

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