Myocarditis (inflammation of the heart muscle)

Background and Assessment

Myocarditis, while rare (1-2% of irAEs), has a high mortality rate (approximately 50%).

Know the risk factors:

- Patients on combination immunotherapy
- Patients with advanced age

Note: However, all patients receiving checkpoint inhibitors can develop myocarditis.

Assess

- Baseline cardiac status (history of CAD, arrhythmias, CHF)
- Consider baseline troponin (T or I) and ECG testing

Look/Listen

- Is the patient experiencing new fatigue or inability to do previous activities?
- Weakness
- New chest pain
- New dyspnea on exertion
- New edema
- Irregular heartrate on exam
- Changes on ECG

Recognize

- Broad range of presenting symptoms, often vague and nonspecific
- May also present as atrial fibrillation, ventricular tachyarrhythmia, or heart block
- Most patients with myocarditis caused by checkpoint inhibitors have an elevated troponin. The majority will also have ECG changes

Myocarditis Workup, Initial Management, and Referral

Initial diagnostic workup:

- Labs: Troponin T or I;* BNP, CK, CRP
- 12-lead ECG
- Echocardiogram

If any initial workup is abnormal:

- Consider urgent admission to the hospital on a monitored bed with a cardiology consultation
- Obtain cardiac MRI
- Hold the checkpoint inhibitor(s)
- High-dose corticosteroid therapy (1000 mg methylpednisolone/day for the first 3 days followed by oral prednisone 1 mg/kg) is usually the first line of therapy in the acute phase

*Ideally, the troponin test type (e.g., T or I) should match the baseline test.

A Note on Triple M

Recognize myocarditis can co-exist with either myositis and/or myasthenia gravis, "Triple M" may present as fatigue, weakness, ocular or bulbar symptoms, respiratory symptoms, and chest pain.

Additional workup for myositis/myasthenia gravis

Myositis initial workup

- CK, aldolase, liver function, CRP
- Necrotizing myopathy panel (myomarker profile -institution dependent)
- Physical exam
- EMG
- Neurology or rheumatology consult

Myasthenia Gravis initial workup

- Anti-AChR antibodies
- EMG
- Paraneoplastic panel
- Neurology consult
- Rule out metastatic disease

AChR = acetylcholine receptor antibody, BNP = B-type natriuretic peptide; CAD = coronary artery disease; CHF = congestive heart failure; CK = creatine kinase; CRP = C-reactive protein; ECG = electrocardiogram; EMG = electromyography; MRI = magnetic resonance imaging.