

# Introduction

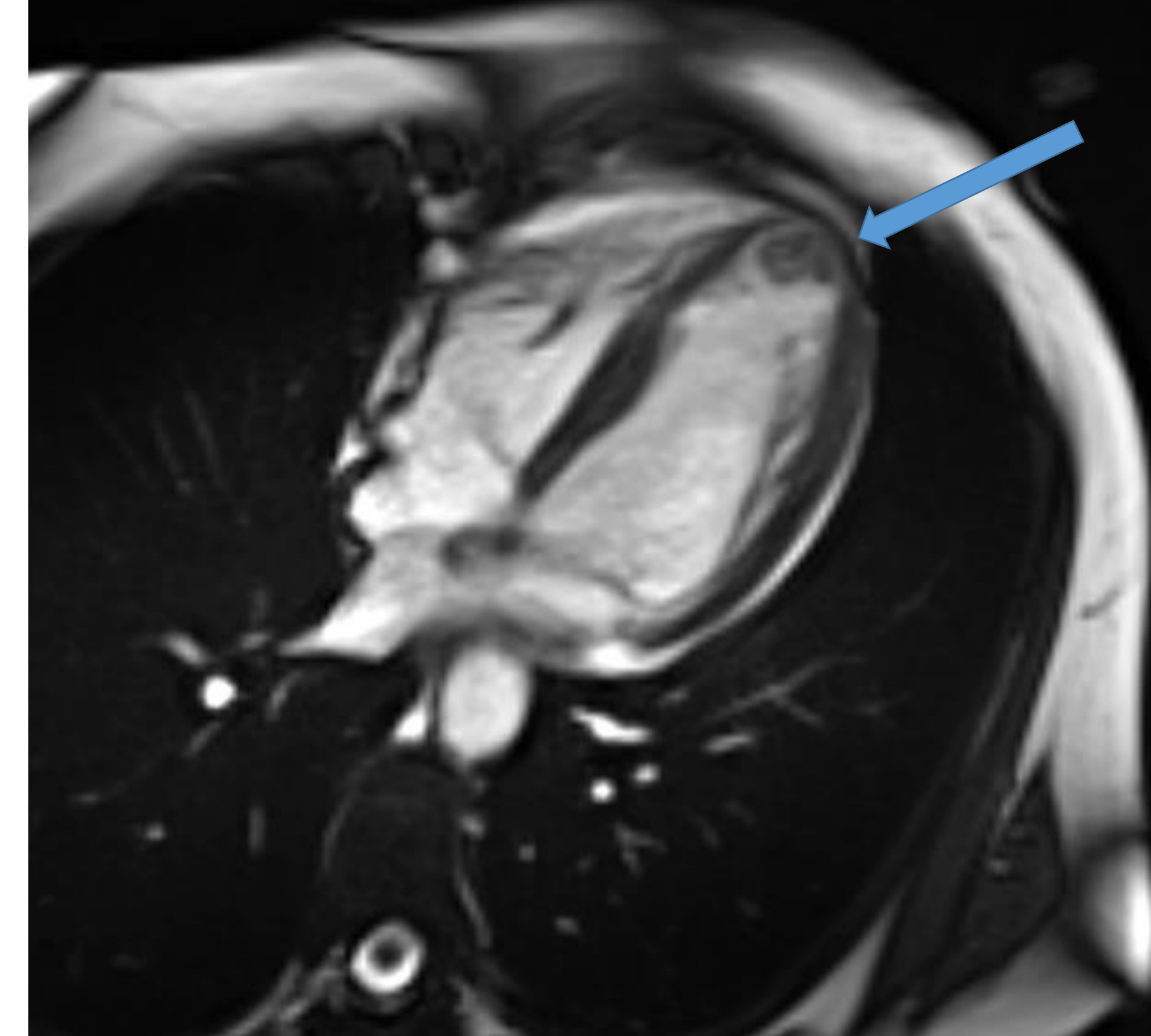
An acute coronary syndrome from vasospasm or plaque rupture accompanying mast cell activation from allergic hypersensitivity or anaphylactoid reactions are referred to as Kounis syndrome. We describe the medically challenging treatment of a 49-year-old female with a history of idiopathic mast cell activation syndrome (MCAS) who presented during an acute cardiac event.

## Proposed criteria for the diagnosis of mast cell activation syndrome

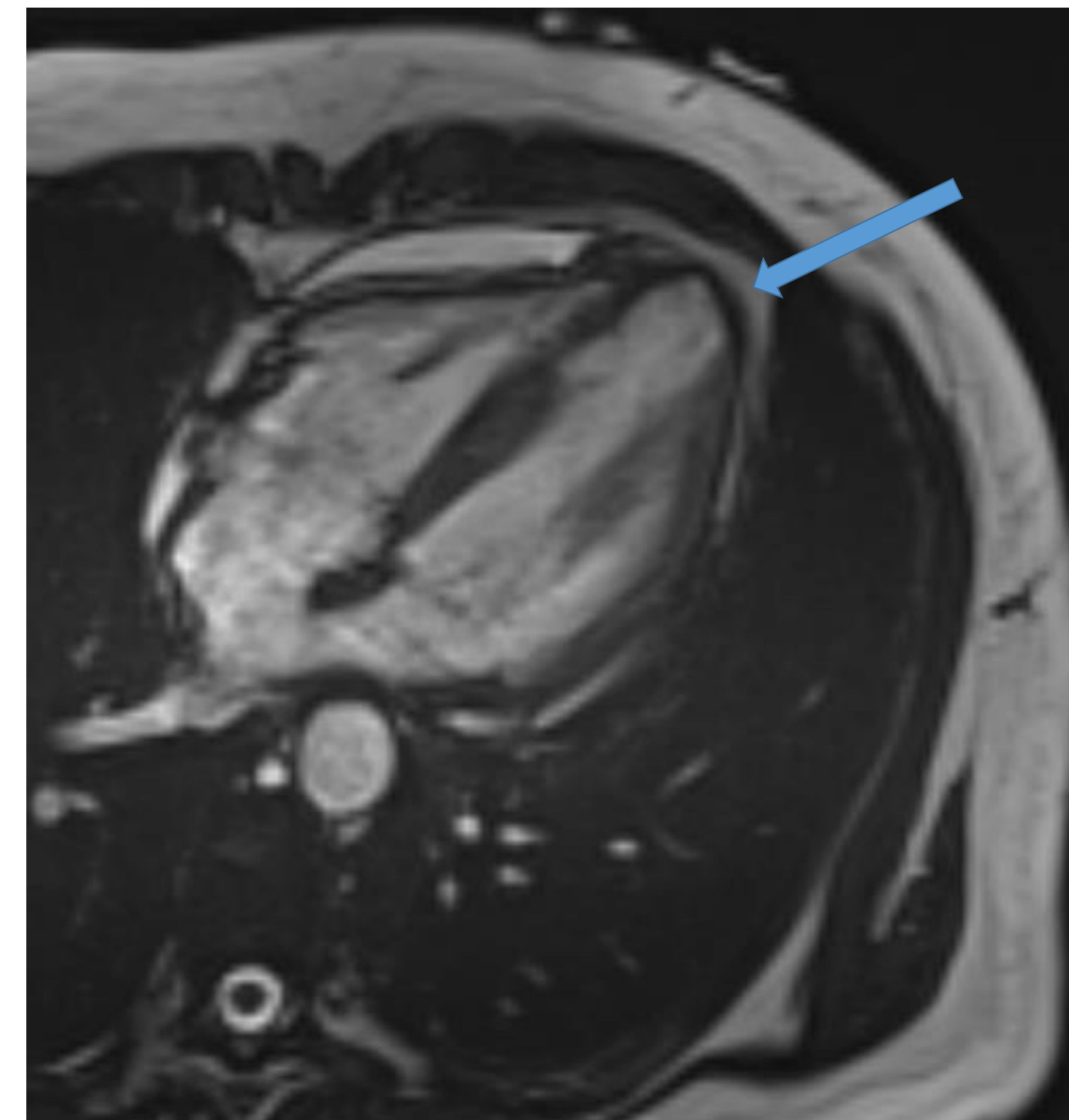
1. Episodic symptoms consistent with mast cell mediator release affecting two or more organ systems evidenced as follows:	
a.	Skin: urticaria, angioedema, flushing
b.	Gastrointestinal: nausea, vomiting, diarrhea, abdominal cramping
c.	Cardiovascular: hypotensive syncope or near syncope, tachycardia
d.	Naso-ocular: conjunctival injection, pruritus, nasal stuffiness
2. A decrease in the frequency or severity; or resolution of symptoms with anti-mediator therapy: H1 and H2 histamine receptor antagonists, anti-leukotriene medications, or mast cell stabilizers.	
3. Evidence of increase in a validated urinary or serum marker of MCAS (preferably with reproducible results obtained during more than one symptomatic episode).	
4. Primary (clonal) and secondary disorders of mast cell activation ruled out.	

MCAS was diagnosed following these proposed diagnostic criteria. This patient had previous recurrent episodes consistent with mast cell mediated release with symptoms ranging from flushing and GI upset, to full systemic anaphylaxis on several different occasions. Patient's symptoms improve and decrease in frequency with daily H1 and H2 histamine receptor antagonists. Tryptase levels during previous episodes have ranged from 13.4 to 14, above her baseline of 7. Bone marrow biopsy was negative for systemic mastocytosis.

\*The opinions or assertions herein are the private views of the authors and are not to be construed as reflecting the views of the Department of the Air Force or the Department of Defense.



Left Ventricular Apical Thrombus – Non-Contrast Cardiac MRI



Resolved Thrombus – 3 Month After Therapy

A 49-year-old female with a history of MCAS presented to the emergency department (ED) with acute shortness of breath. Her initial presentation was however due to her history of MCAS. She was not able to undergo a cardiac MRI. Her cardiac imaging showed a left ventricular apical thrombus with an ejection fraction of 45%.

This case presented a medical emergency. The use of epinephrine could destabilize the patient. The associated increase of heart rate and blood pressure with systemic anaphylaxis had to be managed. The patient received mast cell degranulation. The patient was treated for anticoagulation and platelet inhibition. The patient's desensitization was completed. The patient was discharged on antiplatelet therapy using aspirin and clopidogrel.

This case demonstrates the importance of recognizing and treating MCAS during an acute cardiac event. The patient's symptoms improved and decreased in frequency with daily H1 and H2 histamine receptor antagonists and antiplatelet therapy for any procedures requiring anticoagulation. The patient's left ventricular thrombus with a 45% ejection fraction resolved with 3 months of antiplatelet and anticoagulation therapy.

## References

Weiler CR, Austen KF, Akin C, Barkoff, MS. Mast cell activation syndrome (MCAS) diagnosis and management. *Journal of Allergy and Clinical Immunology*. 2014;134(5):1101-1110.  
Leru, Polliana Mihaela et al. "Mast cell activation syndrome: a review of clinical practice (Review)." *Experimental and Applied Acupuncture* 2017;34(1):1-10.