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Case Report

A Case of Kounis Type I in a Young Woman With Samter's Triad

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ABSTRACT

Kounis syndrome is defined as an acute coronary syndrome triggered by allergic or hypersensitivity reactions resulting in mast cell and platelet activation. Numerous causes have been described, including various drugs, medical conditions, and environmental exposures. Samter's triad consists of nasal polyps, asthma, and aspirin (or nonsteroidal anti-inflammatory drug) sensitivity. We describe a case of Kounis type I in a young woman with Samter's triad who presented with cardiac arrest on 3 occasions. Ergonovine provocation testing established the diagnosis of coronary vasospasm. The patient has derived significant benefit from calcium channel blockers.

RÉSUMÉ

Le syndrome de Kounis se définit comme un syndrome coronarien aigu déclenché par des réactions allergiques ou une hypersensibilité qui entraînent l'activation des mastocytes et des plaquettes. De nombreuses causes expliquent ce phénomène, notamment la prise de divers médicaments, certains troubles médicaux et l'exposition à des facteurs environnementaux. La triade de Samter est un syndrome qui associe la présence de polypes nasaux et d'asthme et une sensibilité à l'aspirine (un anti-inflammatoire non stéroïdien). Nous décrivons un cas du syndrome de Kounis de type I chez une jeune femme atteinte de la triade de Samter qui consulte après avoir subi un arrêt cardiaque à trois reprises. Un test de provocation à l'ergométrine a permis de poser un diagnostic d'angiospasme coronarien. La patiente a réussi à améliorer considérablement son état grâce à la prise d'un inhibiteur calcique.

Kounis syndrome is defined as an acute coronary syndrome triggered by allergic or hypersensitivity reactions resulting in mast cell and platelet activation. Numerous causes have been described, including various drugs, medical conditions, and environmental exposures.¹ The diagnosis of Kounis syndrome requires a high level of suspicion, making it a rarely diagnosed disease.² We describe a case of Kounis type I in a young woman with a history of aspirin-induced asthma who presented with cardiac arrest on 3 occasions.

Case Presentation

A 20 year-old woman was evaluated after 3 episodes of pulseless electrical activity (PEA) over a 7-month period. She had been experiencing worsening asthma and was followed in the severe asthma clinic, where she was managed with fluticasone and salmeterol, salbutamol, and tiotropium inhalers. Her first PEA arrest occurred in the setting of ibuprofen use for headache. She presented with chest tightness, severe

dyspnea, and a troponin level of 24 ng/mL (normal < 0.05 ng/mL). She deteriorated with a PEA arrest, and the electrocardiogram (ECG) showed marked sinus bradycardia at 30 bpm and biphasic T waves in V₁-V₃, which was concerning for ischemia. She was treated for severe asthma exacerbation secondary to nonsteroidal anti-inflammatory drugs (NSAIDs). A diagnosis of Samter's triad was made, which consisted of nasal polyps, asthma, and aspirin (or NSAID) sensitivity.

She had a second PEA arrest 4 months later after ingesting acetaminophen. She had a peak troponin level of 0.67 ng/mL and an ECG demonstrating sinus rhythm. An acetaminophen challenge resulted in severe dyspnea, which resolved with epinephrine. The third PEA arrest occurred after celecoxib use 2 months later, when she presented with dyspnea, chest tightness, and marked sinus bradycardia at 30 bpm with intermittent slow junctional rhythm and recurrent loss of consciousness. The troponin level was 0.21 ng/mL. A single-chamber pacemaker was implanted during that hospitalization.

One month later, she complained of episodes of frequent chest tightness. After a severe episode, she suffered loss of consciousness and had a generalized tonic-clonic seizure. Her troponin level rose to 4.5 ng/mL and her ECG was normal. A transthoracic echocardiogram revealed a left ventricular ejection fraction of 45% with isolated anteroseptal hypokinesis. Three months later, she suffered another troponin rise to

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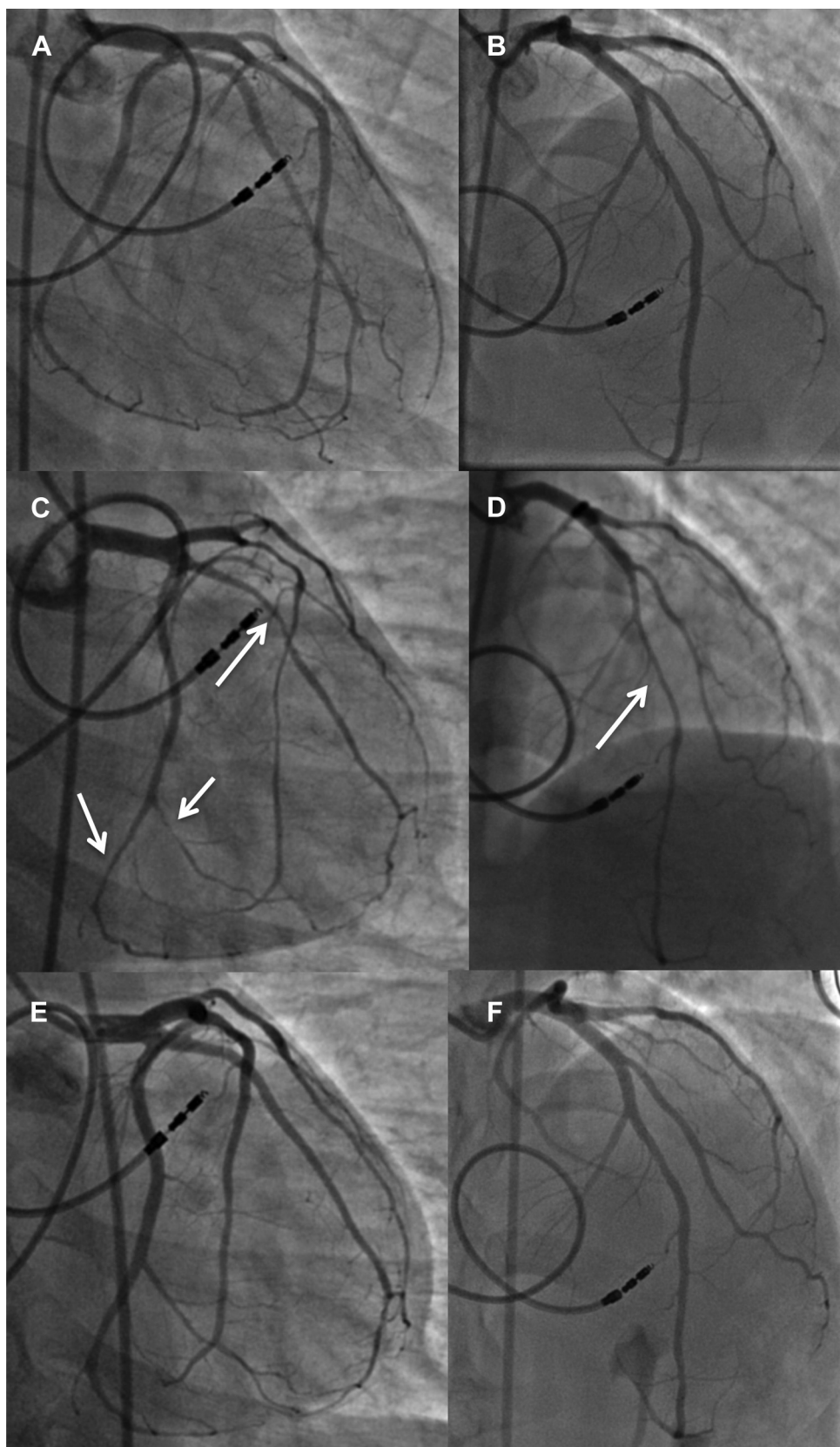


Figure 1. Baseline coronary angiogram showing normal coronary arteries in (A) right anterior oblique (RAO) caudal view and (B) posteroanterior (PA) cranial view. Diffuse coronary vasospasm after ergonovine infusion in (C) RAO caudal and (D) PA cranial views. Complete resolution of vasospasm after nitroglycerin administration in (E) RAO caudal and (F) PA cranial views. **Arrows** indicate areas of stenosis in the ramus, left circumflex, first marginal branch and left anterior descending arteries.

1.5 ng/mL after a prolonged episode of chest tightness relieved with nitroglycerin spray. Coronary provocation testing was performed following a standard protocol consisting of intravenous administration of incremental doses of ergonovine during angiography. Multivessel vasospasm was observed with 0.2 mg of ergonovine, demonstrating 80% stenosis in the ramus, 90% stenosis in the distal left circumflex artery, 90% stenosis in the first marginal branch artery, and 70% stenosis in the mid-left anterior descending (LAD) artery from a baseline of 20% LAD artery stenosis. Intracoronary nitroglycerin completely relieved all stenoses (Fig. 1).

She was administered amlodipine, which was titrated to 5 mg daily, which has significantly improved her chest tightness. She has not had any further PEA arrests.

Discussion

Three types of Kounis syndrome have been described. Type I occurs in patients with normal coronary arteries without coronary artery disease (CAD) risk factors. An acute release of inflammatory mediators results in coronary artery spasm, causing myocardial ischemia or infarction. Type II occurs in patients with pre-existing CAD and involves either spasm or acute plaque rupture. Type III includes coronary artery stent thrombosis and is confirmed with hematoxylin-eosin and Giemsa staining of the aspirated clot, indicating the presence of eosinophils and mast cells.¹

Samter and Beers first described a triad of asthma, aspirin sensitivity, and nasal polyps, which is now known as Samter's triad.³ Aspirin-induced asthma has been associated with type I Kounis syndrome.⁴ Vasospasm is confirmed by the presence of coronary spasm with intracoronary provocation testing

during angiography. A positive response consists of transient > 90% narrowing after ergonovine administration.⁵ Overlapping features between Samter's triad and type I Kounis syndrome are the allergic stimulus, which is typically induced by aspirin or NSAIDs.

We present an unusual case of a young woman with Samter's triad and recurrent PEA arrests secondary to hypoxemia and bradycardia in the setting of bronchospasm, anaphylaxis, and coronary vasospasm with recurrent troponin elevation. Invasive provocation testing demonstrated severe multivessel vasospasm, and she has improved significantly with calcium channel blockers and avoidance of allergens.

Disclosures

The authors have no conflicts of interest to disclose.

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