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ST Elevation Myocardial Infarction as Presenting Feature of C-ANCA Vasculitis: A Case of a Diagnostic Dilemma

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ABSTRACT

Spontaneous coronary artery dissection (SCAD) is a rare cause of acute coronary syndrome (ACS). Although uncommon, it should be included in the differential diagnosis for middle-aged patients without elevated atherosclerotic vascular disease risk or a family history of cardiovascular disease. SCAD is associated with postpartum women; however, reports noting its association with autoimmune disease and vasculopathy in other populations have recently gained prominence. We report a case of a 41-year-old male who was found to have SCAD after presenting with ST segment elevation myocardial infarction in the context of episodic vision loss, and who later underwent work-up for C-ANCA vasculitis and was successfully treated with corticosteroids.

LEARNING POINTS

- SCAD is most common in middle-aged females. However, it can present in male patients, and it should raise suspicion of underlying vasculopathy.
- Eosinophilic vasculitis may mimic parasitic infection.
- C-ANCA vasculitis can be associated with SCAD.

KEYWORDS

Vasculitis, C-ANCA, ST elevation myocardial infarction

CASE PRESENTATION

A 41-year-old Hispanic male, with no reported past medical history, presented to the emergency room with a chief complaint of chest pain. The patient reported a 3-week history of intermittent exertional, retrosternal chest pain associated with nausea and diaphoresis relieved by rest. Moreover, the patient noted that he experienced right temporal headaches with radiation to the frontal and left temporal distributions that occurred multiple times per day. These headaches lasted approximately 20 minutes per episode and were associated with right or bilateral vision loss characterized as diffuse blurriness followed by total blindness. An electrocardiogram revealed ST segment elevations in leads II, III and aVF (*Fig. 1*), warranting emergent cardiac catheterization. A left heart catheterization revealed a distal left anterior descending artery dissection graded as a type 2 spontaneous coronary artery dissection (SCAD) (*Fig. 2*) due to diffuse luminal deformities and TIMI 3 flow indicating proper coronary bed perfusion.

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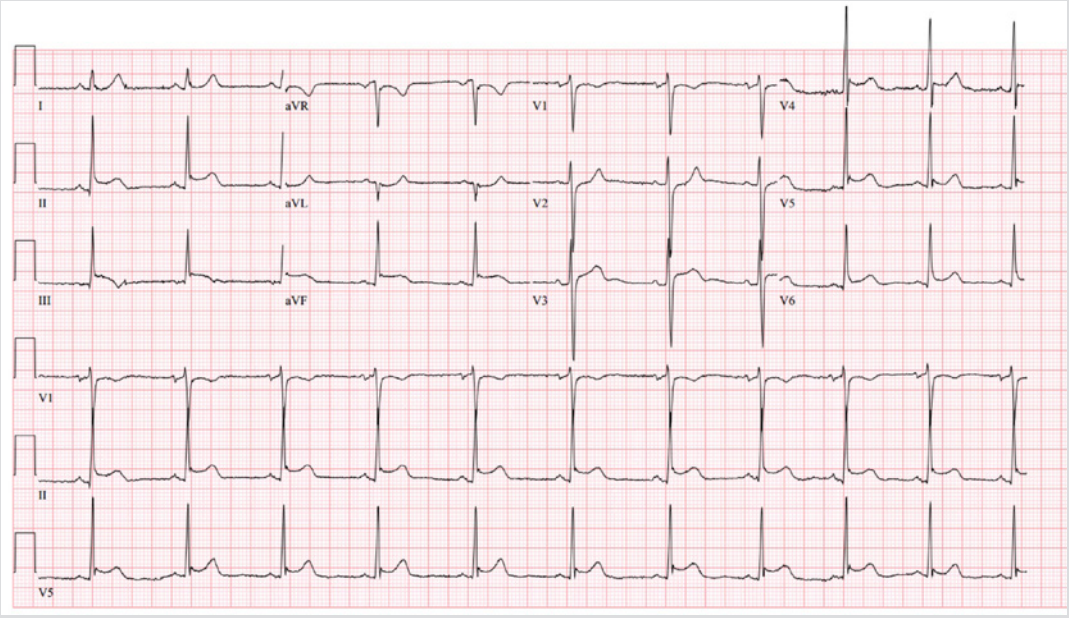

Figure 1. ECG at the time of presentation showing ST elevation at leads II, III and aVF

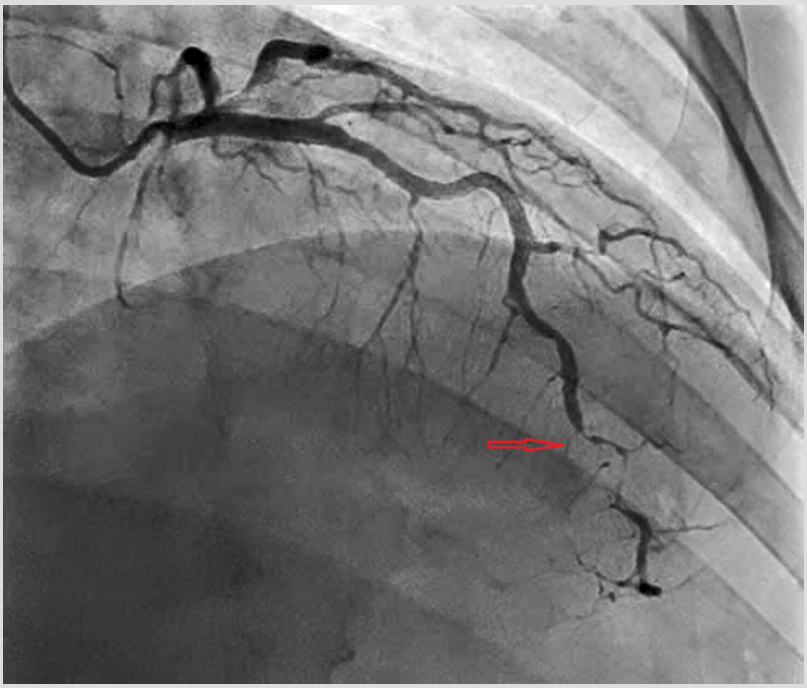

Figure 2. Coronary angiography showing distal left anterior descending artery type 2 spontaneous coronary artery dissection (red arrow)

Based on these findings, no intervention was performed, and the patient was started on medical management with nitroglycerin and heparin. During the hospitalization, his angina resolved. However, he had worsening episodic vision loss, increasing inflammatory markers and an increase in the eosinophil count (1,248 cells/µl, 8% on Day 1 of admission to a peak of 8,382 cells/µl, 33%) per complete blood count. These findings, coupled with fever and headache, prompted a work-up for vasculitides versus infectious aetiology. Negative blood cultures, unremarkable lumbar puncture and unremarkable infectious serological titres made an infectious aetiology less probable. However, the patient received several antimicrobial agents while awaiting further work-up without serologic or clinical improvement. Computerized tomography of the head and neck, as well as an angiography study of the aorta, was conducted to visualize the vasculature, which showed normal structures without focal stenosis and a lack of significant findings of vasculitis. MRI of the brain and orbit revealed bilateral optic neuritis and inflammatory changes of the orbit; however, the patient's retinal examination was unremarkable. An extensive autoimmune

work-up was negative for IgG4, ANA, SSA, SSB, RNP, complement C3 and complement C4. Of note, however, was a positive serine 3 protease marker. At this point, a diagnosis of nonspecific C-ANCA vasculitis was made. A trial of corticosteroids resulted in significant rapid attenuation of eosinophilia and resolution of symptoms. Of note, the patient is a landscaper who frequently travelled to Mexico and had a recent history of camping and wild meat consumption.

DISCUSSION

SCAD is a relatively rare cause of acute coronary syndrome (ACS). It is characterized by a non-atherosclerotic tear in the coronary arteries, which then causes the development of an intramural haematoma within the vessel wall. Subsequent compression of the true lumen may occur, leading to a hindrance to adequate coronary blood flow followed by ACS. The reported incidence of SCAD in patients presenting with ACS varies as established by large-scale studies and is between 0.49% to approximately 2.0% [1, 2].

Attempts to characterize the demographics of patients with SCAD have revealed a significant propensity for SCAD patients to be young to middle-aged postpartum women, with case reports also elucidating possible associations with hormonal therapy, connective tissue disease and high-intensity exercise [3, 4].

While traditionally described in female populations with an emphasis on fibromuscular dysplasia, recent findings have suggested an association of SCAD with vasculopathy. Specifically, it is thought that chronic inflammation of the vasculature leads to weakening of the intimal lining of the vessels, making them highly susceptible to injury. Among these vasculopathies are diseases affecting connective tissue and systemic autoimmune conditions [5]. While the association of connective tissue disease and SCAD has been addressed somewhat, albeit scarcely, in the literature, C-ANCA vasculitis and SCAD association has been documented in a few reports only.

Our case posed a very difficult diagnostic dilemma. The headache, fever, episodic vision loss, subtle optic neuritis and lack of other features typical of vasculitis made an infectious aetiology more likely within the differential diagnosis. The probability of an infectious aetiology was elevated due to the patient's job as a landscaper, with reported frequent exposure to ticks, fleas, mosquitoes and both domesticated and wild animals. Additionally, his travel history and recent camping trip with wild meat consumption put him at an increased risk of parasitic infections. Therefore, the initiation of corticosteroids was delayed until an infectious aetiology was excluded.

It is noteworthy that our patient did not fit a discrete vasculitis designation. Notable differentials for small-vessel vasculitis that promote cardiovascular disease include eosinophilic granulomatosis with polyangiitis, granulomatosis with polyangiitis and microscopic polyangiitis. However, the lack of significant sinus and pulmonary symptoms, and benign findings upon laryngoscopy by otolaryngologists, made eosinophilic granulomatosis with polyangiitis and granulomatosis with polyangiitis less likely. Additionally, microscopic polyangiitis was deemed unlikely due to the lack of cutaneous, musculoskeletal and renal compromise.

Given the uncommon patient demographic, in addition to the lack of reports in the literature characterizing C-ANCA vasculitides with SCAD, awareness of encounters such as these hold value in informing clinicians and provide utility in guiding the prospective work-up of vasculopathy in those with SCAD without a delineated origin.

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