Case Report

Fulminant Vasculitis Complicated by ST-Elevation Myocardial Infarction and Stroke

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Acute coronary syndrome is a rare complication of vasculitis. We present a case of fulminant medium-vessel vasculitis, most likely PAN, complicated by STEMI and stroke, that was successfully treated with percutaneous revascularization, high-quality stroke care, and immunosuppression. This case highlights the importance of prompt diagnosis and treatment of vasculitis and the recognition of coronary and cerebral ischemia as potentially serious complications.

1. Introduction

Systemic vasculitides are rarely associated with acute myocardial infarction or stroke. Prompt diagnosis and treatment is critical given the increased morbidity and mortality of systemic vasculitis with cardiovascular involvement [1].

2. Case Report

A 30-year-old woman with a one-year history of recurring painful lower extremity skin lesions presented with acute headache, neck pain, and right arm weakness with paresthesia. She was tachycardic but afebrile and normotensive. CT angiography of the head and neck revealed high-grade stenosis of the internal carotids. Subsequently, the patient reported acute chest pain and dyspnea. ECG revealed sinus tachycardia, anterior ST elevation, and reciprocal inferior depression (Figure 1). Labs revealed elevated troponin, serum lactate, and white blood cell count (Table 1). She was given aspirin and intravenous bivalirudin and underwent emergent cardiac catheterization that revealed an acute occlusive thrombus of the proximal left anterior descending (LAD) artery and estimated Fick cardiac index 1.9 L/min/m² (Figure 1, Table 1). Aspiration thrombectomy was performed, and a 4.5 × 30 mm drug-eluting stent was successfully deployed in the proximal LAD. The patient was started on a norepinephrine infusion and transferred to our institution with concern for fulminant medium-vessel vasculitis complicated by STEMI, cardiogenic shock, and stroke.

Upon arrival to our hospital, exam demonstrated expressive aphasia, left upper quadrantanopia, and right-sided hemiparesis. Bilateral upper extremity pulses were normal, femoral pulses were reduced, and dorsalis pedis and posterior tibial pulses were absent. A pulmonary arterial catheter was placed (Table 1). Neurology was consulted and urgent head and neck CT angiography revealed high-grade occlusions of the bilateral internal carotid arteries and stigmata of left middle cerebral artery infarct (Figure 1). Brain MRI confirmed evolving left frontal and parietal lobe infarcts (Figure 1). Echocardiogram demonstrated severe left ventricular systolic dysfunction with regional wall motion abnormalities: anterior,
septal, and apical. CT imaging demonstrated abnormal wall thickening of the thoracic aorta, common iliac arteries, and renal arteries, as well as occlusion of the inferior mesenteric and infratibial arteries. Rheumatology was consulted and a broad serologic evaluation was initiated (Table 1).

2.1. Management. The patient was treated with IV corticosteroids and cyclophosphamide. Shock physiology, likely secondary to myocardial stunning, rapidly resolved. Systemic anticoagulation was discontinued after 48 hours following the determination that arterial occlusions were attributable to vasculitis rather than thrombophilia. Dual antiplatelet therapy with clopidogrel and aspirin was continued.

2.2. Follow-Up. On hospital day 10, the patient was transferred to acute rehabilitation for poststroke care. She was prescribed prednisone, cyclophosphamide, maintenance dual antiplatelet therapy, and a heart failure regimen. Rheumatology recommended outpatient adenosine deaminase 2 (ADA2) genetic testing. Three months after discharge, she was doing well on maintenance immunosuppression and following with cardiology for chronic heart failure.

3. Discussion

Cardiac involvement in vasculitis is not uncommon and can include cardiomyopathy, pericarditis, valvular disease, arrhythmia, aortic dissection, and coronary complications such as aneurysm, stenosis, thrombosis, or rupture [1]. However, STEMI is an uncommon initial presentation of vasculitis. This report highlights the diagnosis and management of fulminant medium-vessel vasculitis, with features of polyarteritis nodosa (PAN) or adenosine deaminase 2 deficiency (DADA2), presenting as STEMI and acute stroke.

PAN is a systemic medium-vessel necrotizing vasculitis that most frequently involves the skin, kidneys, and peripheral nervous system and is diagnosed by a combination of clinical, angiographic, and pathologic findings [2]. Cardiovascular involvement occurs in only 5-22% of patients and carries a two to threefold higher mortality compared to those without coronary involvement [3]. Coronary involvement in PAN is angiographically characterized by alternating areas of stenosis or occlusion with aneurysm in a “beads on a string” pattern [1, 2]. Treatment involves aggressive immunosuppression, typically with corticosteroids and cyclophosphamide [2]. DADA2 is an autoinflammatory disorder first described in 2014 that clinically resembles PAN and is characterized by vasculitis, dysregulated immune function, and hematologic abnormalities [4, 5]. Pathophysiologically, lack of ADA2 leads to proinflammatory endothelial dysfunction, predisposing patients to systemic vasculitis and recurrent strokes, often beginning in childhood [6]. Significant clinical overlap and lack of commercial enzyme testing makes differentiating ADA2 deficiency from PAN challenging, though genetic testing for CECR1 mutations can aid in diagnosis. While there is no clearly preferred immunosuppressive therapy for DADA2, tumor necrosis factor inhibitors may be particularly effective [5].
### Table 1: Clinical, laboratory, and echocardiographic data.

| Vitals                          | Day 1 | Day 2 | Day 4 |
|---------------------------------|-------|-------|-------|
| Temperature, °C                 | 35.6  | 36.2  | 36.3  |
| Heart rate, beats/min           | 150   | 143   | 98    |
| Blood pressure, mmHg            | 120/105 | 123/87 | 113/73 |
| Respiratory rate, breaths/min   | 25    | 30    | 22    |
| SpO₂, %                         | 100   | 98    | 99    |
| Body mass index, kg/m²          | 32    | 32    | 32    |

| Laboratories                    |       |       |       |
|---------------------------------|-------|-------|-------|
| Troponin I, ng/L                | 7.0   | —     | —     |
| Hs-troponin, ng/L               | —     | 206,452 | 174,327 |
| NT-proBNP, ng/mL                | —     | 1,059 | —     |
| White blood cells, 10⁶/L         | 53    | 25    | 16    |
| Hemoglobin, g/dL                | 16    | 13    | 10    |
| Platelets, 10⁹/L                | 460   | 364   | 254   |
| Creatinine, mg/dL               | 0.60  | 0.80  | 0.50  |
| Aspartate transaminase, IU/L     | 1,478 | 533   | 113   |
| Alanine aminotransferase, IU/L   | 158   | 120   | 53    |
| International normalized ratio  | 5.1   | 2.0   | 1.1   |
| Lactate, mmol/L                 | 6.2   | 3.6   | 1.8   |
| Erythrocyte sedimentation rate, mm/hr | N/A | 58 | — |
| C-reactive protein, mg/dL       | 2.9   | 125   | —     |
| Hemoglobin A1C, %               | 5.5   | 5.4   | —     |
| Total cholesterol, mg/dL        | 193   | 137   | —     |
| Triglycerides, mg/dL            | 224   | 146   | —     |
| HDL cholesterol, mg/dL          | 44    | 33    | —     |
| LDL cholesterol, mg/dL          | 104   | 79    | —     |

| Echocardiogram                  |       |       |       |
|---------------------------------|-------|-------|-------|
| LVEDV, mL                       | 115   |       | 174   |
| LVESV, mL                       | 89    |       | 110   |
| LVEF, %                         | 23    |       | 37    |
| Left atrial volume, mL/m²       | 15.5  |       | 15.3  |
| RVSP, mmHg                      | —     |       | 44.6  |
| Valvular heart disease          | None  |       | None  |

| Cardiac hemodynamics            |       |       |       |
|---------------------------------|-------|-------|-------|
| RA pressure, mmHg               | 3     | 5     | 5     |
| PA mean pressure, mmHg          | 26    | 25    | 27    |
| PCW pressure, mmHg              | —     | 15    | —     |
| Cardiac output, L/min           | 4.1   | 3.8   | 5.7   |
| Cardiac index, L/min/m²         | 1.9   | 1.7   | 2.6   |

| Infectious studies              |       |       |       |
|---------------------------------|-------|-------|-------|
| HIV                             | Negative |     |       |
| Hepatitis B DNA PCR             | Negative |     |       |
| Hepatitis C                     | Negative |     |       |
| Blood cultures                  | Negative |     |       |

| Rheumatologic studies           |       |       |       |
|---------------------------------|-------|-------|-------|
| ANA titer                       |       | 1:320, speckled |       |
| Anti-ds-DNAb                     |       | Negative |       |
Takayasu’s arteritis (TA) is a large-vessel vasculitis primarily involving the aorta and its major branches that usually affects young women with preexisting cardiac abnormalities [1]. Coronary artery stenosis and occlusions usually occur at the ostia because of the extension of inflammation-induced intimal proliferation and subsequent fibrotic retraction from the ascending aorta [7]. Treatment involves high-dose immunosuppression. TA was felt to be less likely in this patient given the predominantly medium-vessel involvement encompassing multiple vascular territories beyond the aorta.

The patient’s clinical presentation, imaging, and laboratory findings were inconsistent with alternative diagnoses such as Behcet’s disease, giant cell arteritis, or other systemic rheumatologic conditions.

4. Conclusions

Acute coronary syndrome is a rare complication of vasculitis. Our patient presented with fulminant medium-vessel vasculitis, most likely PAN, complicated by STEMI and stroke, and was successfully treated with percutaneous revascularization, high-quality stroke care, and immunosuppression. This case highlights the importance of prompt diagnosis and treatment of vasculitis and the recognition of coronary and cerebral ischemia as potentially serious complications.

Data Availability

Data supporting the conclusion may be made available via communication with the corresponding author.

Additional Points

Learning Objectives. (1) Recognize vasculitis as a rare etiology of STEMI. (2) Recognize vasculitis as an uncommon etiology of stroke. (3) Emphasize necessity of early recognition of vasculitis and prompt immunosuppression.

Conflicts of Interest

The authors declare no conflict of interest.

References

[1] L. H. Silveira, “Cardiovascular manifestations of systemic vasculitides,” Current Rheumatology Reports, vol. 22, no. 10, p. 72, 2020.
[2] A. Hočevar, M. Tomšič, and P. K. Perdan, “Clinical approach to diagnosis and therapy of polyarteritis nodosa,” Current Rheumatology Reports, vol. 23, no. 3, p. 14, 2021.
[3] D. P. Misra and S. N. Shenoy, “Cardiac involvement in primary systemic vasculitis and potential drug therapies to reduce cardiovascular risk,” Rheumatology International, vol. 37, no. 1, pp. 151–167, 2017.
[4] P. Navon Elkan, S. B. Pierce, R. Segel et al., “Mutant adenosine deaminase 2 in a polyarteritis nodosa vasculopathy,” The New England Journal of Medicine, vol. 370, no. 10, pp. 921–931, 2014.
[5] A. Human and C. Pagnoux, “Diagnosis and management of ADA2 deficient polyarteritis nodosa,” International Journal of Rheumatic Diseases, vol. 22, Suppl 1, pp. 69–77, 2019.
[6] Q. Zhou, D. Yang, A. K. Ombrello et al., "Early-onset stroke and vasculopathy associated with mutations in ADA2," The New England Journal of Medicine, vol. 370, no. 10, pp. 911–920, 2014.
[7] M. Endo, Y. Tomizawa, H. Nishida et al., "Angiographic findings and surgical treatments of coronary artery involvement in Takayasu arteritis," The Journal of Thoracic and Cardiovascular Surgery, vol. 125, no. 3, pp. 570–577, 2003.