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Dissect The Case

Kaitlyn L. Buzard DO
Lehigh Valley Health Network, Kaitlyn.Buzard@lvhn.org

Kaitlyn Dalsey DO
Lehigh Valley Health Network, kaitlyn.dalsey@lvhn.org

Lisa E.A. Dwyer-Joyce MD
Lehigh Valley Health Network, lisa.dwyer-joyce@lvh.com

Nicole Chiappetta DO
Lehigh Valley Health Network, nicole.chiappetta@lvhn.org

James Ross MD, FACP
Lehigh Valley Health Network, James.Ross@lvhn.org

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Published In/Presented At

Buzard, K.L., Dalsey, K., Dwyer-Joyce, L., Chiappetta, N.C., & Ross, J. (2022). *Dissect The Case*. Poster presented at Lehigh Valley Health Network, Allentown, PA.

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Dissect The Case

Kaitlyn L. Buzard, DO,¹ Kaitlyn Dalsey, DO,¹ Lisa Dwyer-Joyce, MD,² Nicole C. Chiappetta, DO,¹ and James Ross, MD¹

¹Department of Rheumatology, ²Department of Pathology, Lehigh Valley Health Network, Allentown, PA

Introduction

Vasculitis: inflammation of blood vessels, typically characterized by size of vessel

MAJOR CATEGORIES:

Large-vessel vasculitis	Medium-vessel vasculitis	Small-vessel vasculitis
Takayasu arteritis Giant cell arteritis	Polyarteritis nodosa Kawasaki disease	ANCA-associated vasculitis Immune complex small-vessel vasculitis

Case Presentation

- 86-year-old-female with a past medical history of HTN, HLD, CKD3, hypothyroidism, and former tobacco use who presented to the hospital with chest pain, back pain, and jaw pain
- Patient was hemodynamically stable and physical exam was unremarkable except for right eye cornea opacification
- STAT imaging revealed type A aortic dissection
- Cardiothoracic surgery was consulted, and patient was taken emergently to the OR for repair and replacement of the ascending aorta and transverse aortic arch with hemiarch technique
- Surgery was without complications and patient was extubated on post-op day #1

Results

- Initial lab work with normal CBC/CMP, normal ESR, and elevated CRP (23.5 mg/L)
- Rheumatological Serologies: +ANA (1:80 cytoplasmic, 1:160 homogenous), Indeterminate Quantiferon Gold, and SPEP with IgM Kappa MGUS. Otherwise, serologies were negative including dsDNA, SSA, SSB, RNP, Smith, SCL-70, RF, CCP, ANCAs, ACE, Vitamin D 1,25, Cryoglobulins, aPLs, IgG4, RPR, Lyme, Hepatitis Panel, and UA
- Histopathology of the aorta revealed acute on chronic focally necrotizing vasculitis consistent with aortitis and intramural dissecting hemorrhage

Diagnosis and Treatment

ADDITIONAL HISTORY OBTAINED:

- Suspected scalp tenderness and hip/shoulder girdle pain and stiffness 3 months prior
- Associated constitutional symptoms including fatigue, weight loss, and decreased appetite
- No headache, vision changes, or jaw claudication

TREATMENT:

- IV Solumedrol 60 mg daily x 3 days
- Transitioned to Prednisone 1 mg/kg (60 mg daily) with prolonged steroid taper
- Later started on IV Actemra 6 mg/kg monthly

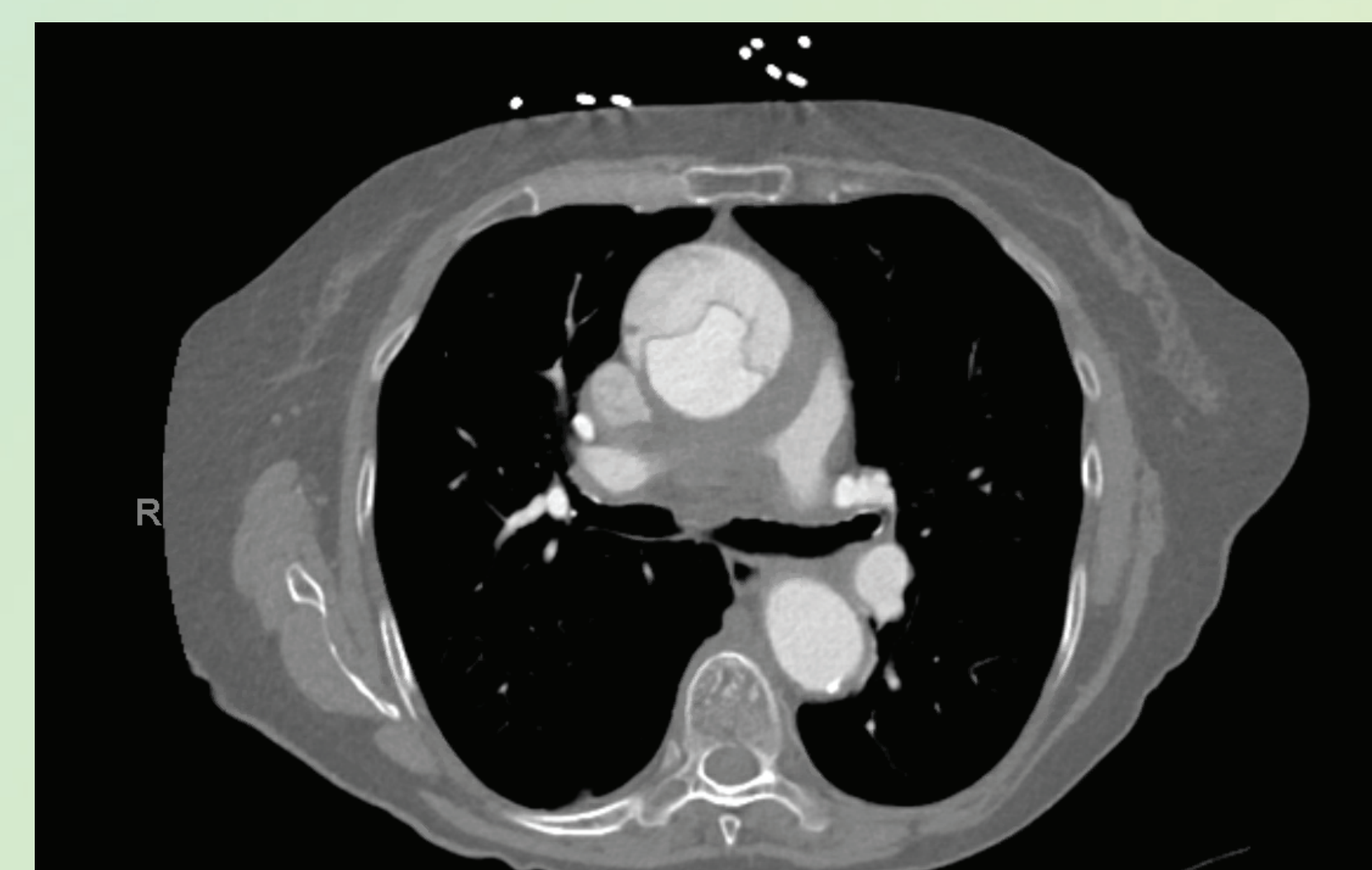
FOLLOW UP:

- Normalization of inflammatory markers (ESR 1, CRP <3.0 mg/L)
- CT C/A/P: Stable thoracic aortic aneurysm with no evidence of leak

Discussion

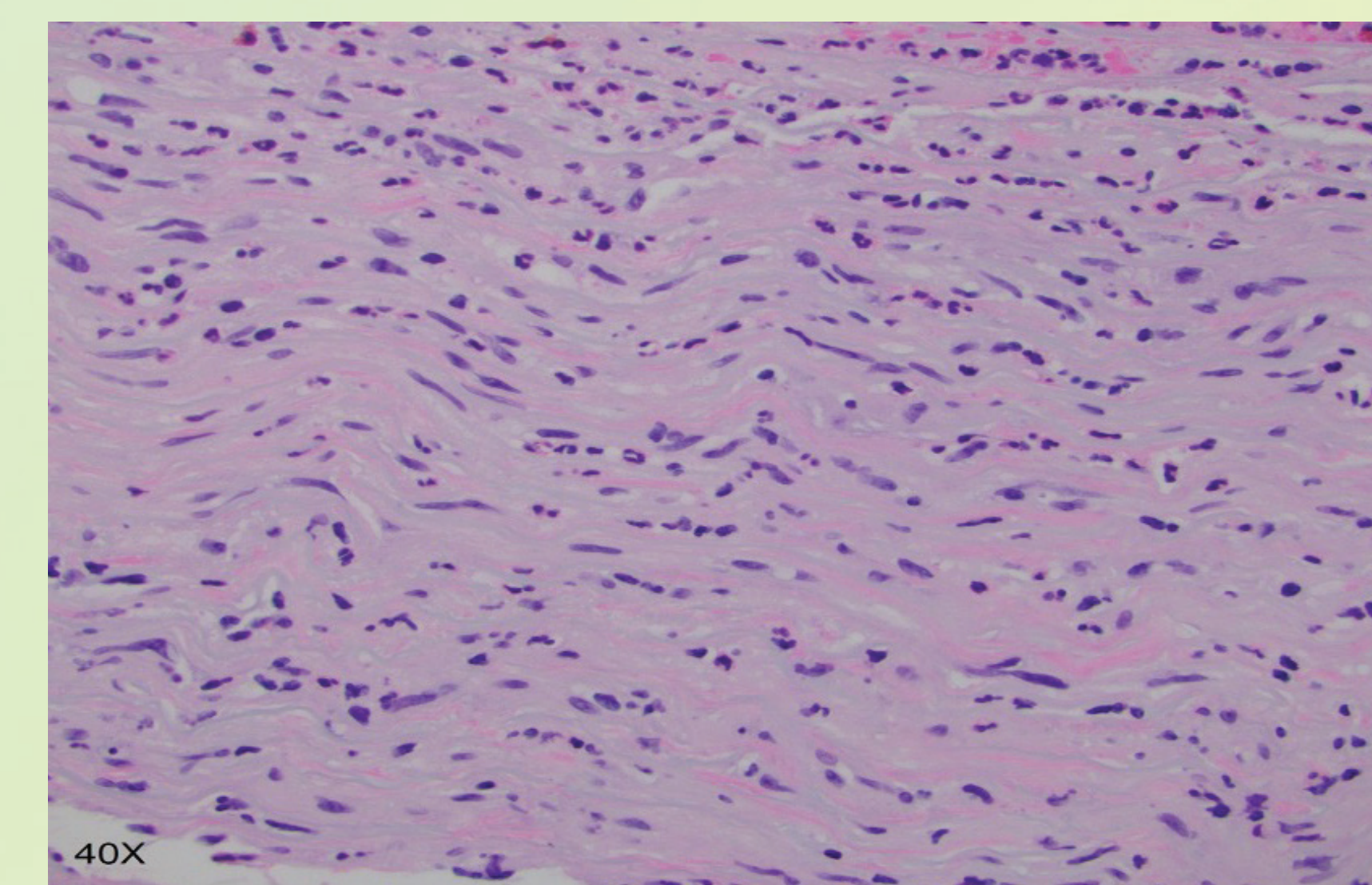
- This is a case of delayed diagnosis in which the patient presented with a life-threatening complication of the most common systemic vasculitis
- Aortic aneurysm is a more common complication (10-20%)
- Aortic dissection:
 - Occurs 1-6% of cases
 - Can occur early or late in disease course
 - Decreased survival when compared to GCA patients without aortic aneurysm/dissection
- Further research should be considered regarding incidence of aortic dissection with and without history of an aortic aneurysm due to the increased mortality and to help guide surveillance recommendations

IMAGING

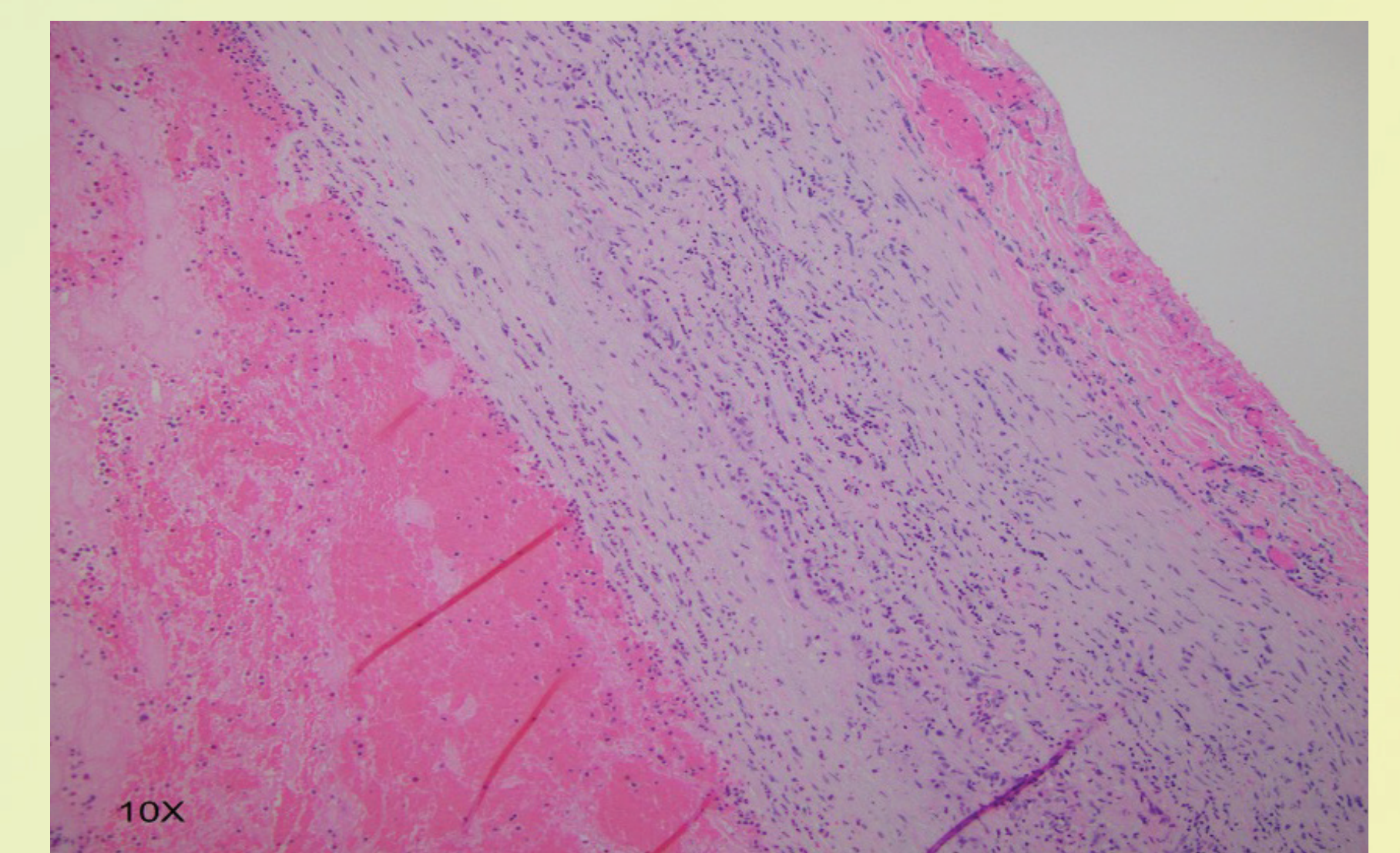


CT Chest Dissection Protocol:
Type A aortic dissection. Postcontrast images demonstrate a type A aortic dissection. The dissection flap terminates proximal to the common trunk of the right and left common carotid artery

TEMPORAL ARTERY BIOPSY



High power, lymphohistiocytic and neutrophilic inflammatory infiltrate



Elastic stain demonstrating disruption elastic fibrils

Photographs courtesy of HNL Lab Medicine - Pathology Department

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