



DEPARTMENT OF PATHOLOGY

Short Report in Pathology

Autopsy

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History:

A 36-year-old female presented to the emergency department with cardiac arrest. She was placed on extracorporeal membrane oxygenation (ECMO). However, her status progressed from ventricular rhythm with a pulse to ventricular rhythm without a pulse. In view of a family history of aortic dissection, a CT scan of the chest was performed which was suggestive of type A Aortic dissection extending from the aortic root to the proximal aortic arch. Mediastinal exploration was conducted but no evidence of aortic dissection was found. The patient expired due to cardiac arrest and an autopsy was conducted to ascertain the cause of the death.

Gross Images:

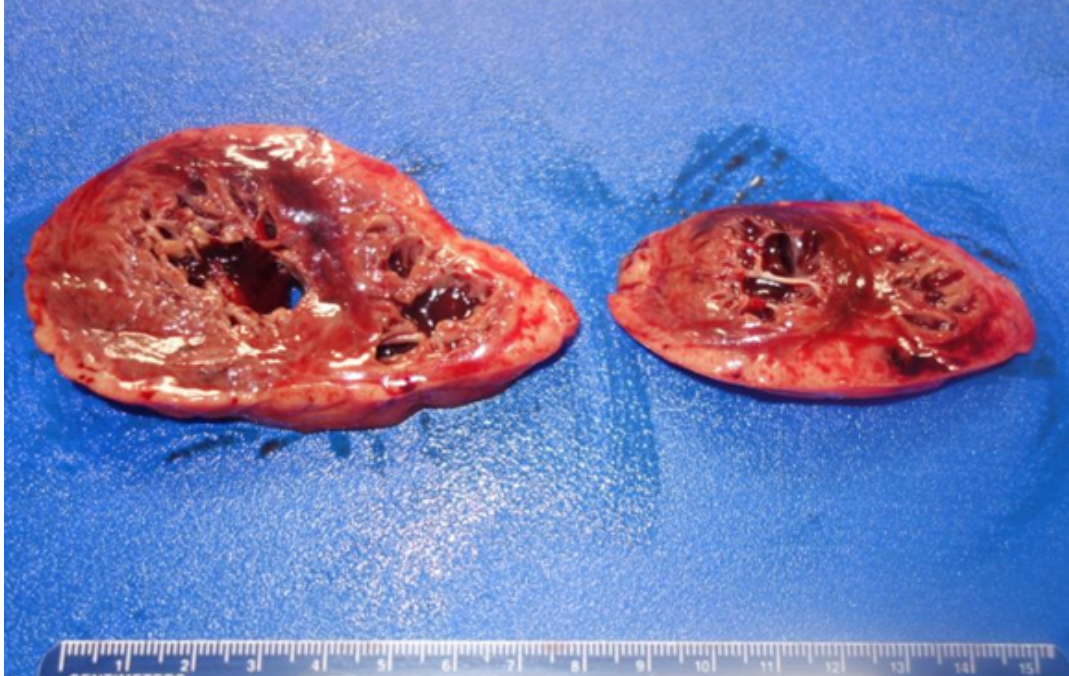


Figure 1: Gross examination of the heart reveals red-brown hemorrhagic areas (white arrow) in the wall of the left ventricle and interventricular septum, suggestive of acute myocardial infarction.

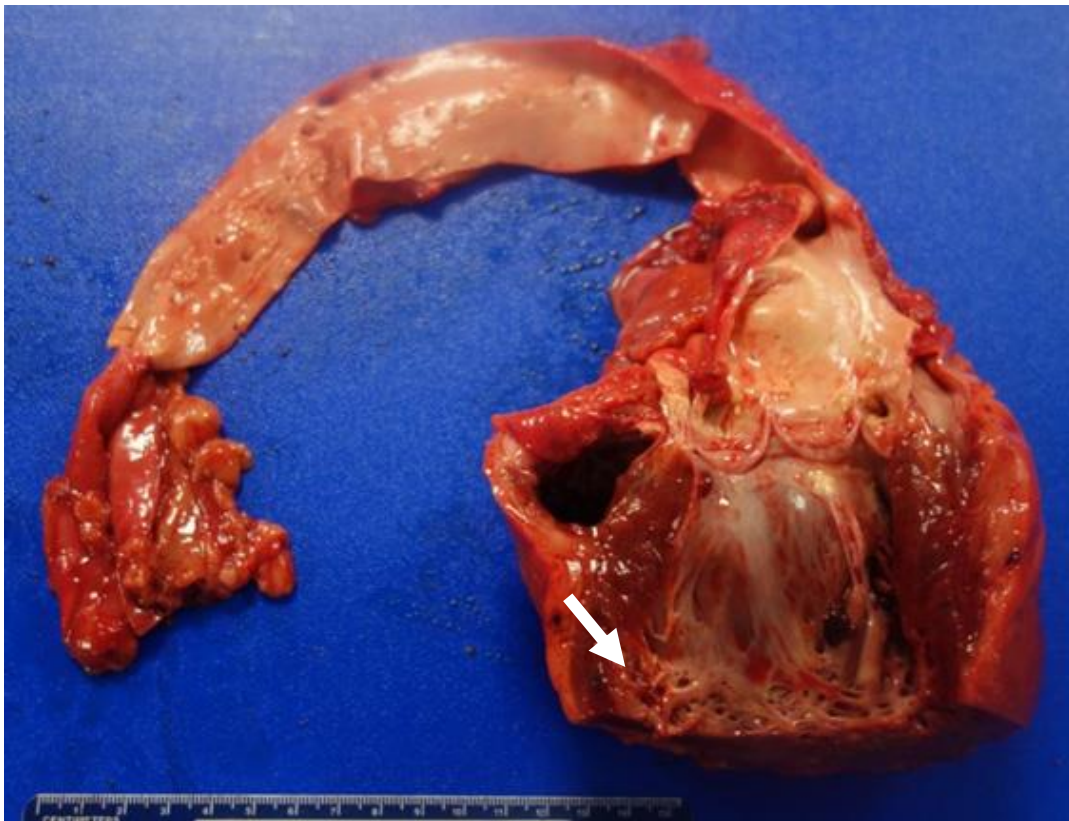


Figure 2: The dissected heart shows a large hemorrhagic area (arrow) which represents extravasated blood (blood clot) around the thrombosed left anterior descending artery.

Microscopic Images:

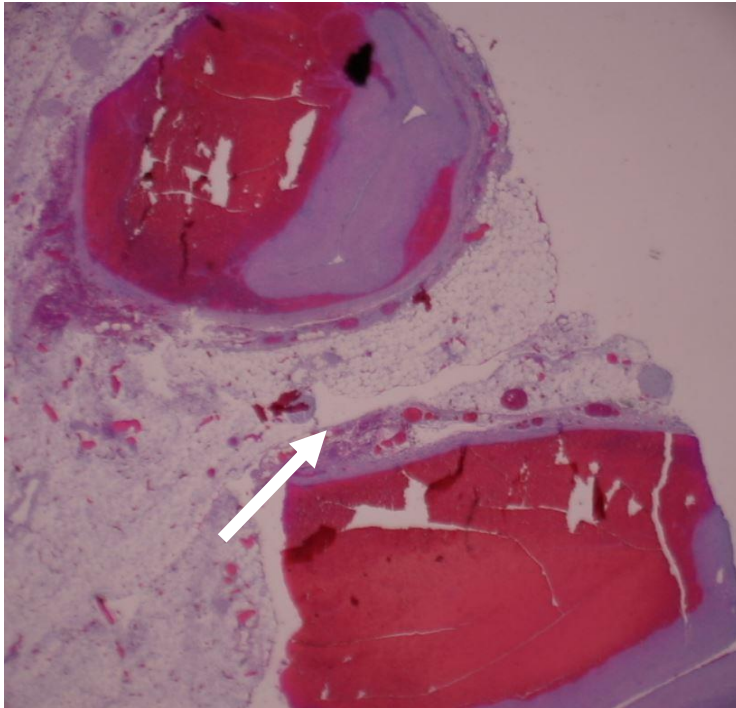


Figure 3: Section of left anterior descending artery showing the dissection of the tunica media as characterized by extravasation of blood between the tunica media and adventitia layers (Hematoxylin and Eosin stain, 200x).

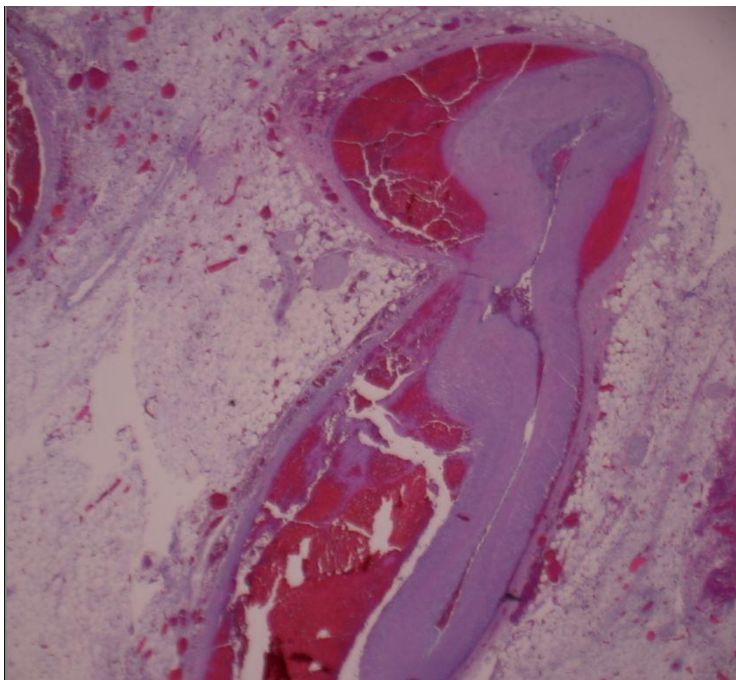


Figure 4: Section from more distal left anterior descending artery showing dissection of blood between media and adventitia layers (Hematoxylin and Eosin stain, 200x).

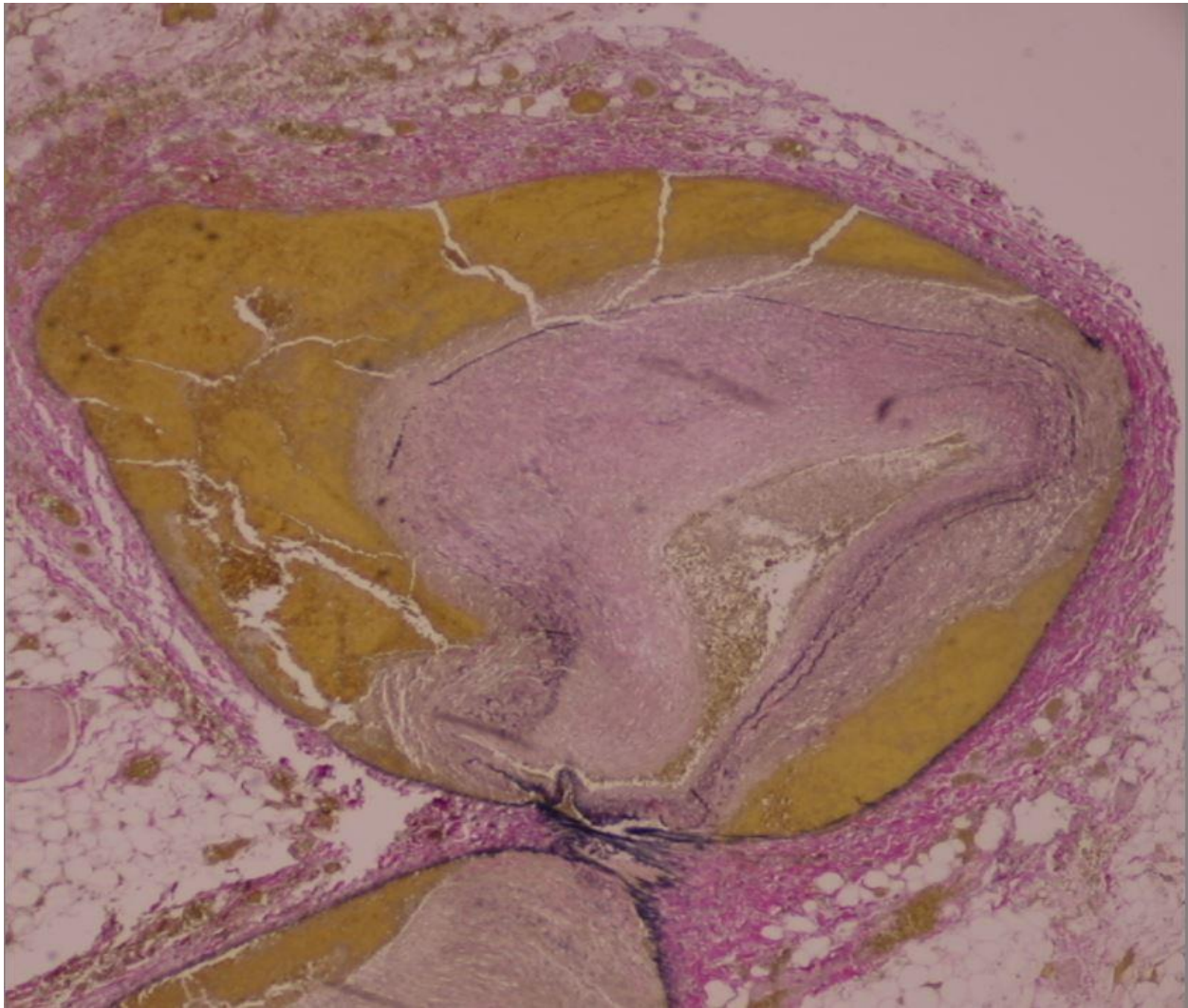


Figure 5: Cross-section from left anterior descending artery showing thickened tunica intima (>2 fold thicker than tunica media) consistent with fibromuscular dysplasia (Elastin stain, 200x).

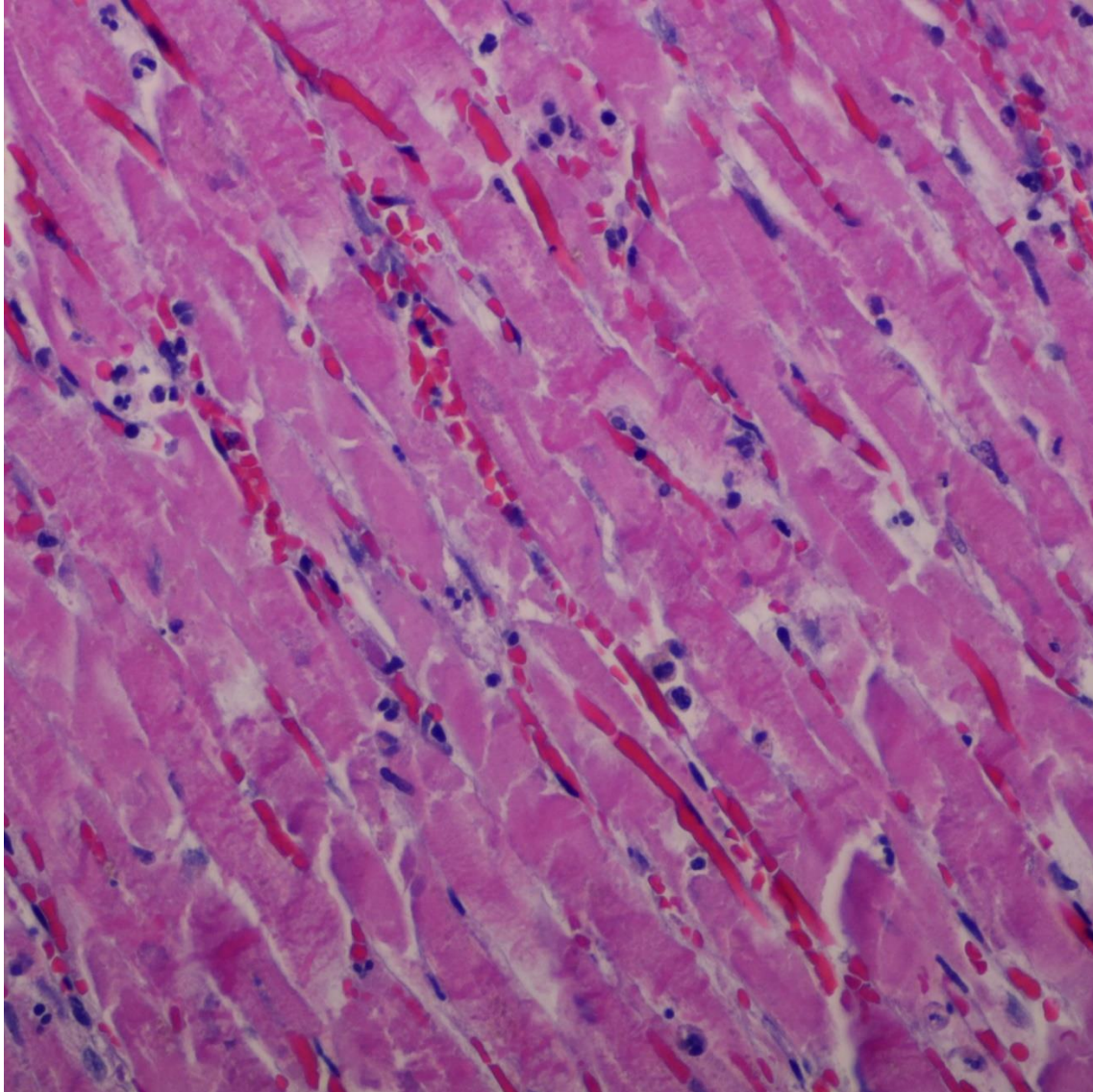


Figure 6: Section from left ventricle showing cardiac myofibers without nuclei infiltrated by neutrophils and congested microvasculature, consistent with infarction (Hematoxylin and Eosin stain, 400x).

Diagnosis:

Spontaneous coronary artery dissection secondary to fibromuscular dysplasia leading to acute myocardial infarction and sudden cardiac death.

Discussion:

Spontaneous coronary artery dissection (SCAD) is an exceedingly rare but potentially life-threatening cause of acute coronary syndromes, including acute myocardial infarction that can lead to cardiogenic shock and sudden death. This condition predominantly affects female, with the left anterior descending artery being the most commonly affected. A number of risk factors

have been identified in association with SCAD, including pregnancy, peripartum and postpartum period, connective tissue disorders like Ehlers-Danlos syndrome, systemic lupus erythematosus, and fibromuscular dysplasia.

Fibromuscular dysplasia (FMD) affecting the coronary arteries is a relatively uncommon yet clinically significant condition that may present as acute coronary syndrome, left ventricular dysfunction, or even sudden cardiac death. It is most likely that subjects with FMD carry a genetic predisposition, as 7% to 11% of first-degree relatives exhibit signs of FMD. Consequently, clinicians should maintain a high index of suspicion for SCAD in young females presenting with acute myocardial infarction in the absence of conventional risk factors for acute coronary artery disease.

While angiography is the standard for diagnostic evaluation, it may fall short in detecting subtle cases of underlying SCAD if the involvement of coronary arteries is subtle. In such substances, the integration of new imaging modalities, such as intravascular ultrasound (IVUS) and optical coherent tomography (OCT), can provide valuable assistance in establishing a definitive diagnosis of SCAD.

References:

1. Farooq A, Amjad W, Bajwa AUR, Yasin H, Ali R, Pervaiz M. Fibromuscular Dysplasia with Spontaneous Coronary Artery Disease Presenting as Acute Myocardial Infarction. *Cureus*. 2017 May;23(5):e1268.doi:10.7759/cureus.1268. PMID: 28652952; PMCID: PMC5481181.
2. Spontaneous coronary artery dissection: prevalence of predisposing conditions including fibromuscular dysplasia in a tertiary center cohort. Saw J, Ricci D, Starovoytov A, et al. *JACC Cardiovasc Interv*. 2013;6:44–52.
3. Coronary artery manifestations of fibromuscular dysplasia. Michelis KC, Olin JW, Kadian-Dodov D, et al. *J Am Coll Cardiol*. 2014;64:1033–1046.