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A Case Report Of Spontaneous Coronary Artery Dissection in a Man With Ehlers–Danlos Syndrome

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ABSTRACT-

Background: Spontaneous coronary artery dissection (SCAD), as a medical emergency, represents one of the non-atherosclerotic causes of an acute coronary syndrome (ACS). It often occurs in young and middle-aged females and is a rarity among male patients. Yet, it is easily misdiagnosed or missed even though it has one of the highest in-hospital mortality rates.

Case summary: Here, we present a young male patient admitted to the emergency department of our hospital due to a complaint of acute chest pain. During his hospitalization, we utilized several tools, including imaging modalities, genetic analyses, and clinical strategies, to ensure a proper diagnosis and management of the patient. The results indicated that the patient suffered from SCAD, as well as vascular Ehlers–Danlos syndrome (vEDS). Unfortunately, the patient died of SCAD-related sudden cardiac death (SCD) on the ninth day before the DNA analysis results were obtained. Despite a global effort and huge progress in the clinical characterization of SCAD, as well as patients' assessments, its pathophysiology remains poorly understood, with a significant recurrence risk and no specific disease-modifying therapy.

Conclusion: Vascular Ehlers–Danlos syndrome, as an inherited connective tissue disorder characterized by congenital connective tissue dysplasia, is a rare and particularly challenging monogenetic disease. It can cause life-threatening changes, including arterial dissections and ruptures, and lead to early death due to COL3A1 pathogenic variants. It is also a rare cause of SCAD. Currently, the gold standard for SCAD diagnosis is coronary angiography (CAG).

KEYWORDS- Spontaneous coronary artery dissection, Ehlers–Danlos syndrome, congenital connective tissue dysplasia, sudden cardiac death