The Journal of Emergency Medicine, Vol. ■, No. ■, pp. 1–11, 2016

0736-4679/$ - see front matter

http://dx.doi.org/10.1016/j.jemermed.2015.10.044

Clinical Review

CERVICAL ARTERY DISSECTIONS: A REVIEW

Jennifer J. Robertson, MD, MSED* and Alex Koyfman, MD†

*Department of Emergency Medicine, Emory University, Atlanta, Georgia and †Parkland Hospital, University of Texas-Southwestern, Dallas, Texas

Reprint Address: Jennifer J. Robertson, MD, MSED, 531 Asbury Circle, Annex Building, Suite N340 Atlanta, GA 30322

□ **Abstract—Background: Cervical artery dissection (CeAD) is an infrequent, yet potentially devastating, cause of stroke. While uncommon, CeAD is important for emergency physicians to quickly diagnose and treat because of the potential for cerebral ischemia, stroke, blindness, or death. To our knowledge, no review articles in the emergency medicine literature have been published on CeAD. A literature search of MEDLINE/PubMed, Embase, and other major abstracts in the English language was performed for the following terms: cervical artery, vertebral artery, and carotid artery dissection. The search included all titles from January 1, 2010 to February 28, 2015 and other relevant articles. Objectives: We sought to review the epidemiology, pathophysiology, risk factors, and clinical presentation for extracranial CeAD in the adult population, explore recent research on diagnosing this disorder, evaluate the most current research on treatment options, and summarize the prognosis of CeAD. Discussion: CeAD is an uncommon but important cause of stroke in the young that is likely caused by multifactorial processes. The diagnosis should be considered in those with underlying risk factors, a remote history of minor trauma, and concerning signs and symptoms. The condition should be pursued via magnetic resonance imaging or computed tomography angiography. Treatment should be aimed at preventing additional complications, including recurrent stroke or transient ischemic attack, with antiplatelets, anticoagulants, or even endovascular or surgical therapy. Conclusion: Overall, the prognosis of patients with CeAD is good, with relatively low death rates. However, the diagnosis should not be missed, because treatment may help prevent worsening or persistent ischemia, recurrent dissection, and death.** © **2016 Elsevier Inc. All rights reserved.**

□ **Keywords—cervical artery dissection; internal carotid artery dissection; vertebral artery dissection**

INTRODUCTION

The cervical arteries comprise both the internal carotid and vertebral arteries. Dissection of one or both can be asymptomatic but can also lead to cerebral ischemia, stroke, blindness, subarachnoid hemorrhage, or death (1–5). While thrombotic stroke is typically a disease of the elderly, cervical artery dissection (CeAD) causes stroke in young and middle-aged patients (2). Only 1% to 2% of all ischemic strokes are caused by CeAD, but in younger patients, CeAD accounts for 10% to 25% of strokes (2). CeAD is a multifactorial process that is not well understood but is likely related to underlying risk factors, such as genetic predisposition and previous trauma (2,6,7).

CeADs are considered to be extracranial or intracranial, but extracranial CeADs are more common (8,9). CeADs are further classified as spontaneous or traumatic, but in reality, many patients with "spontaneous" dissections have sustained mild trauma (6,10–12). The bulk of the recent literature focuses on

RECEIVED: 17 May 2015; FINAL SUBMISSION RECEIVED: 22 September 2015;
ACCEPTED: 17 October 2015

adults >18 years of age with spontaneous extracranial CeAD and those who have sustained minor trauma (13–17). Major trauma, such as blunt aortic trauma extending into the cervical arteries, and purely intracranial dissections have different mechanisms and symptoms and are managed differently (2). Given the most recent relevant research, the focus of this review will remain on adults with spontaneous extracranial CeAD and those who have sustained minor trauma leading to CeAD.

Incidence

Approximately 1% to 2% of all ischemic strokes are caused by CeAD, but they produce up to 25% of ischemic strokes in young individuals (2,8,16,18,19). The estimated annual incidence of CeAD is 2.6 to 5 patients per 100,000 patients per year (16,19,20). However, the overall incidence of CeAD is likely underestimated because some patients have no symptoms, subtle symptoms, or an uncertain onset of symptoms (1,21). Reports on the incidence of vertebral artery dissection (VAD) versus internal carotid artery dissection (ICAD) also differ, although several studies show ICAD to be more common than VAD (16,22–24). While rare, case reports do show that bilateral and multiple simultaneous dissections can occur (4,16,19,23). Interestingly, CeAD tends to occur more often in autumn and winter (25).

The mean age of occurrence of CeAD is approximately 44 years of age, and it is rare beyond 65 years of age (7,16,23). It occurs most frequently in the fifth decade of life (2,26). Observational studies show CeAD to be slightly more common in men, but overall, there are no large differences in the incidence of CeAD between genders (8,26). VAD is more common in younger individuals, especially women, and ICAD is more prevalent in older men (4,26,27). It is important to mention that most epidemiologic studies on CeAD are observational and based on European and American populations (16,19). Therefore, it should be kept in mind that the incidence of CeAD is estimated and that patterns may be different in other populations (2).

Pathophysiology and Risk Factors

CeAD is characterized as a hematoma within the wall of the internal carotid or vertebral arteries (2,8). Initially, a tear occurs in the artery where blood enters the arterial wall under pressure. Separation of the wall layers causes a false lumen where blood leaks into the vessel wall, forming the hematoma. This causes an even narrower lumen, potentially leading to occlusion (28,29). The hematoma and the thrombus can lead to cerebral thromboembolism, decreased blood flow, and subsequent ischemic stroke (8,30). It may also lead to mass effect on surrounding structures, causing disorders such as Horner's syndrome (29,31). While hypoperfusion is a potential cause of ischemia, it has been found that thromboembolism causes the majority of ischemic strokes in patients with CeAD (31).

It remains somewhat unclear what actually causes CeAD. Some reported risk factors include genetic predisposition, minor trauma, migraine headaches, pregnancy and postpartum, previous infection or inflammation, connective tissue disease, strangulation, and hypertension (2,4,6,32–42). Many authors postulate that the condition is likely multifactorial, with both genetic predisposition and minor trauma playing more significant roles in the development of CeAD (2,7,8,11,21). Several case reports describe CeAD in patients sustaining minor trauma, such as whiplash, roller coaster rides, self-contained underwater apparatus diving, stretching, chiropractic manipulations, sports and exercise, turbulent flights, and even dental procedures (43–60). Manual strangulation can cause CeAD, but it is uncommon (40,41). Clinicians should pursue the diagnosis of CeAD in strangulation victims only when patients are unconsciousness or have unilateral neurologic signs (41). However, patients may not develop neurologic symptoms for up to 36 hours, so clinicians should observe any victim of strangulation who has a suspected CeAD (41). This includes patients with voice changes, loss of consciousness, physical evidence of neck trauma, and neurologic examination abnormalities (41).

It is estimated that minor trauma plays a role in approximately 40% of cases of spontaneous extracranial CeAD (6). Despite the suspected role of minor trauma, it is not typical for minor traumas, such as whiplash or sudden head movements, to cause CeAD in most individuals. Therefore, it is hypothesized that patients who sustain a CeAD with or without minor trauma likely have an underlying arteriopathy, inflammatory process, or structural instability of the arteries that leads to dissection (1,2,4,7,28,37). In fact, a 2011 study by Volker et al. shows biopsy-proven structural differences in the arterial walls of patients with spontaneous CeAD and in those patients who have sustained major trauma (28). A 2015 study by Saba et al. also shows a positive association with dissection and kinking and coiling of the internal carotid artery, which suggests an underlying predisposition (21).

Because of the limited understanding of the pathophysiologic mechanisms of CeAD, the Cervical Artery Dissection and Ischemic Stroke Patients (CADISP) consortium was established in 2009 to further study the genetics, risk factors, clinical presentation, and outcome predictors of CeAD (13). CADISP is the largest known international multicenter collection of CeAD patients, and also includes age-, sex-, and country-matched non-CeAD ischemic stroke and healthy patients for