Eagle Syndrome and Sudden and Unexpected Death: Forensic Point of View about One Case

Clément R and Barrios L
Laboratoire de médecine Légale, Faculté de médecine, 1 Rue Gaston Veil, 44 035 NANTES Cedex, France

Abstract

The causes of sudden death in young adults under the age of 35 are mainly due to cardiovascular issues. We report the case of a 25 year-old man who died suddenly at the wheel of his car, very likely while he was reversing. The autopsy only discovered bilateral ossification of the stylohyoid ligaments. The microscopic and toxicological investigations showed strictly normal results. The cause of death and the circumstances involved in it are discussed under the framework of these ligament anomalies, called “Eagle syndrome”. Indeed, the circumstances involved in the death led to envisaging an abrupt pressure from the vasculonervous structures of the neck on the ossification of the stylohyoid ligaments, during rotation of the neck carried out by the victim due to the accident while reversing.

A cardio inhibitory reflex is referred to as the cause of death, but abrupt pressure from the carotid artery cannot be adopted as a contributing factor due to the absence of histological lesions. Moreover, unlike the two cases reported in the forensic literature, and the clinical description of a cerebrovascular accident related to Eagle syndrome, the pressure had to be exercised on the nervus vagus. It is then possible to attribute the cause of death to overstimulation of it whilst in a state of stress.

Keywords: Eagle syndrome; Autopsy; Sudden death; Cardioinhibitory reflex

Introduction

Sudden death is defined by the abrupt and unexpected death of a subject in good health that occurs within one hour [1]. The primary causes in people under the age of 35 are mainly due to cardiovascular issues [2]. Hypertrophic cardiomyopathies and cases of arrhythmogenic dysplasia of the right ventricle are the primary causes of this. Other extracardiac causes are known, such as pulmonary conditions (pulmonary embolism, asthma), neurological conditions (intracerebral hemorrhage, epilepsy), and abdominal conditions (hemorrhagic process). The case reported envisages an exceptional cause of unexpected sudden death linked to Eagle syndrome, resulting from histological anomalies of the stylohyoid ligament complex. But the following observation also leads to the debated issue of death due to a cardioinhibitory reflex caused by neck pressure. And in the case at hand, due to the combination of a neck movement that led to pressure by the neck's vascular-nervous elements on these rigid internal intracervical structures that result in ossification of these ligaments.

Case Report

A 25 year-old man was discovered in his vehicle at a car park near his home. A group of young people sounded the alarm right at the end of the night as they were intrigued by the fact that this vehicle's lights were still on, even though it had stopped. Initial observations made in situ noted that the subject was sitting in the driver's seat, with his seatbelt on, and with his head rotated laterally to the right. The vehicle was in reverse gear and a rear wheel had crossed the metal boundary marker delineating the car park area. The police investigation
Figure 1: CT findings; elongated bilateral ossification of the stylohyoid (Sh)

Figure 2: dissection of left stylohyoid ligament (V: vagal nerve; C: bifurcation of left carotid; E: left ossified stylohyoid ligament)

Figure 3: left calcified stylohyoid ligament; extracted from the neck

Figure 4: Right calcified stylohyoid ligament; extracted from the neck
Discussion

Eagle syndrome was established subsequent to the description of it provided by Watt W. Eagle [3]. The stylohyoid chain consists of the styloid bone process, the small horn of the hyoid bone, and the stylohyoid ligament, which joins the two structures. Eagle syndrome is subdivided into two clinical entities depending on the different symptomatological characteristics noted subsequent to gathering 250 cases [4]. The "standard" syndrome is characterized by odynophagia, dysphagia, cervicalgia, and craniofacial pain. The "carotid" syndrome entails faintness, feelings of intoxication, and even cerebrovascular accidents. From an anatomical perspective, the cause of Eagle syndrome can be elongation of the styloid process (above and beyond 40 millimeters), ossification of the stylohyoid ligament, or both. In this area of the anatomy, the stylohyoid chain is in contact with the jugular vein, the internal carotid artery, the nervus vagus, and the hypoglossal nerve [5].

The epidemiological data provide an estimated frequency in the general population in 4% of cases. Some indicate a frequency of 1.4% [6] in young subjects (1 to 18 years of age), while others indicate a frequency of 30% for the elderly [7]. Only 4 to nearly 10% of the subjects afflicted present with symptoms. This symptomatic group is characterized by the preponderance of women and an age of over 50 [6,7]. Conversely, an autopsy study relating to 1,215 cases [8] discovered a frequency of 0.9% with men predominating (7/11). Two cases were discovered in subjects under the age of 30. Nine cases were bilateral.

This case presented leads to several questions being raised. The profile – a 25 year-old man – does not match the traditional descriptions reported in the literature. Likewise, according to the investigations, he did not present with any symptomatology prior to death that could match one of the two types of Eagle syndrome. That was concluded in the light of reported investigations with close relation. But in Eagle's syndrome, the symptoms range from mild discomfort to acute neurologic. The symptoms may be confused with episodic physical disorders. It is possible that the person could not complain about these non-specific symptoms.

Two cases were published concerning the deaths of people who were discovered in autopsies to have experienced changes in the stylohyoid chain [9,10]. The Indian case [9] relates to faintness caused by Eagle syndrome that was the source of fatal trauma resulting from the victim falling. There were pre-existing faintness-type symptoms prior to the incident. Another case published by a Sri Lankan team [10], indicated a symptomatic history in a 39 year-old woman. In this latter case, episodes of faintness were noted, as well as headaches. The cause of death in this publication is linked to Eagle syndrome which, in the authors' view, is characterized by a bilateral elongation of the two styloid processes measuring 3 and 3.7 cm and prolonged by a partial calcification of the stylohyoid ligaments. Due to the pressure exerted on the carotid bifurcation by these rigid structures, cardiac arrhythmia may occur. An exaggeration of this inhibition reflex may cause loss of consciousness and be complicated by cardiac arrest. The authors adopted this physiopathological process because, upon being taken under medical supervision, it was noted that the subject was experiencing hypotension and an irregular pulse. In support of their demonstration, flexion, extension and rotation movements were carried out during the autopsy operations that determined a significant compression of the bone structures on the carotid glomeruli. In order to certify this cause of death, the theory involving hyperactivity of the reflex by the carotid sinus was adopted. And yet recent publications contest the use of this physiopathological mechanism for deaths resulting from a fatal neck pressure without any proof other than the absence of macroscopic, microscopic and toxicological elements [11,12]. A recent article refutes reflex death caused by fatal pressure in the neck without any histological lesion of the glomerulus [11]. A meta-analysis was carried out by a Swiss team [12] that excluded cardioinhibitory reflex death without preexisting factors that may be the simultaneous presence of toxins, cardiac conditions, or states of physical or psychological excitation.

In the case presented, the death of this young man occurred under circumstances suggestive of rotation and abrupt extension of the neck. As a matter of fact, the observations in situ assume that he was reversing, and abruptly crossing the metal structure prompted him to look back at what was happening. And then death occurred. This rotation and extension of the neck, which was probably vigorous, may have provoked a forced, exaggerated bearing down by the vascular and lateral nerve elements of the neck on the rigid structure of the left stylohyoid chain. Although carotid bifurcation cannot be the cause of an exaggeration of the cardioinhibitory reflex due to the absence of any histological lesion, it is anatomically possible that the exaggerated
pressure may have caused hyperactivity of the nervus vagus, which ties in with descriptions of reflex deaths [13]. The anatomical structures close to an elongation of the styloid process concern the nervus vagus [14]. For this, another stimulation factor is required, and several publications demonstrate vagal reflex deaths during a state of stress [15,16]. Likewise, neck pressure due to ossification of the stylohyoid ligaments applying to the glomeruli, it seems, can be ruled out. Several cases in the literature describe vascular accidents caused by cases of Eagle syndrome involving the extracranial internal carotid artery [17,18]. A study on cadavers even demonstrated that Eagle syndrome compresses the internal carotid artery and not the glomerulus [19]. All of these anatomical, surgical and imaging data constitute additional factors for ruling out hyper reactivity of the cardio inhibitory reflex due to overstimulation of the carotid glomerulus. Although in the case reported, it is impossible to determine the psychodynamic state of the victim at the time of death, it is not impossible to consider that the exaggerated reversing of the vehicle could be evidence of stress. This Hypothesis of a reflex death due to Eagle syndrome with excessive irritation of the nervus vagus, does however hinge on several prognostications.

In conclusion, although the theory involving cardioinhibitory death does require histological lesions of the carotid bifurcation as well as predisposition factors [11,12], it does now turn out to be essential to take samples from these areas when a fatal neck pressure is envisaged that may involve a hanging, strangulation, or death by a direct blow to the neck. It would also doubtless be very useful to take samples from the nervivagi in order to implicate another reflex cardiac death process in these situations. Because although the theory involving an inhibitory cardiac reflex is accepted, practical application of it does however remain controversial [20].

References