

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

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### 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 glomeruli 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 nervusvagus. 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

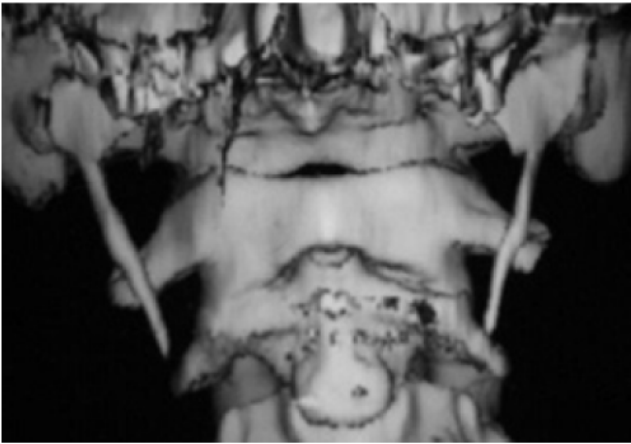
### Introduction

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 intra-cervical 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

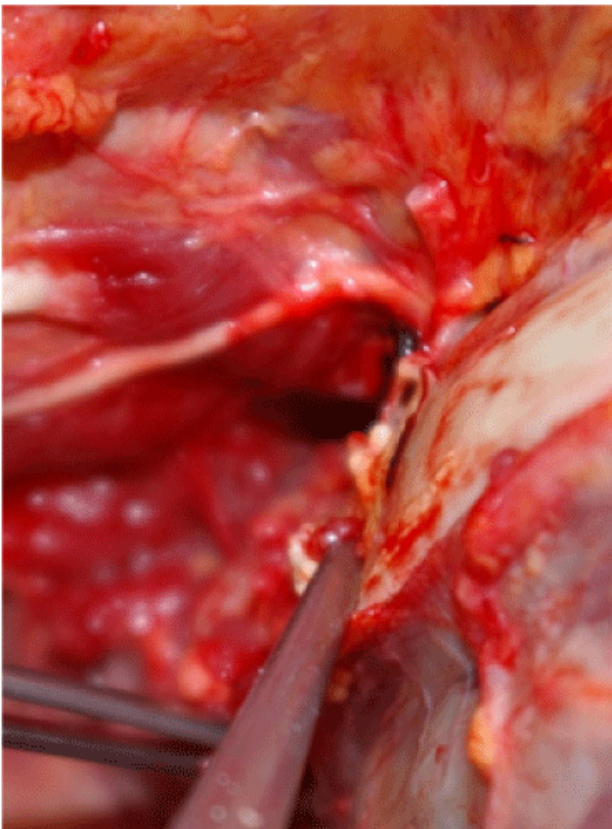
established that this young man, who had been discovered dead at 6.30 am, had been expected at his place of work at 5 am. According to his family and his doctor, he had no previous medical or surgical history, was not taking any treatments, and did not take any drugs. He simply had a proclivity for being much stressed. An autopsy was carried out. This young man was 1.75 m tall and weighed 73 kilos (Figure 1). No lesions were found during the external examination. There were no signs of cyanosis or petechial hemorrhage. The internal examination found that his heart weighed 345 grams. His macroscopic examination was normal. The lungs featured neither any congestion nor edema, with the left lung weighing 280 grams and the right lung weighing 320 grams. The macroscopic and histological examinations turned out to be normal. There were no intrinsic subtle cardiac complaints. The examinations of the meninges and the brain were normal. To the contrary, dissection of the neck was hindered by two rigid structures. Two calcified processes of the two stylohyoid ligaments were dissected, measuring 5 cm on the left (Figures 2 and 3) and 3 cm on the right (Figure 4). The external and internal walls of the carotid arteries were normal macroscopically, as were the carotid glomeruli (Figure 5). Microscopic exploration of them proved to be normal. The stylohyoid ligaments are the seat of the calcified bone and bone marrow structures (Figure 6). The carotid arteries were normal. The toxicological results did not highlight any recent or chronic consumption of toxic substances. Upon completion of all of these macroscopic, microscopic and toxicological explorations, it was concluded that the cause and circumstances of the death were natural and a sudden death triggered by a reflex caused by Eagle syndrome is suggested.



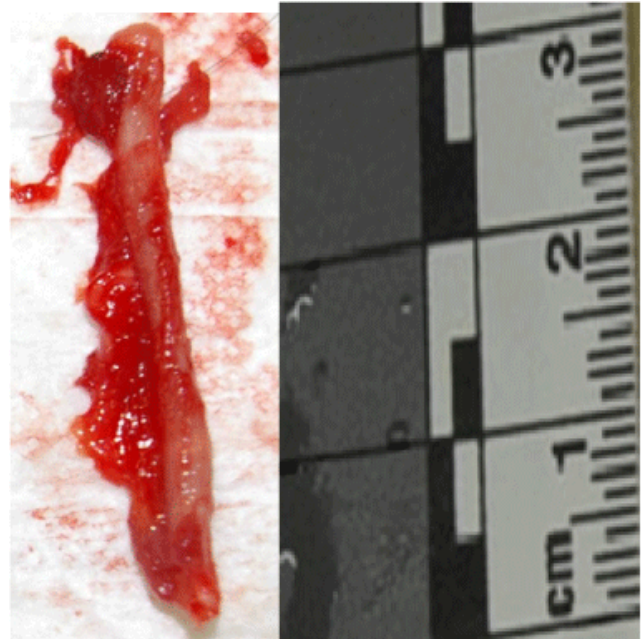
**Figure 1:** CT findings; elongated bilateral ossification of the stylohyoid (Sh)



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



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



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



Figure 5: Left carotid artery and its bifurcation

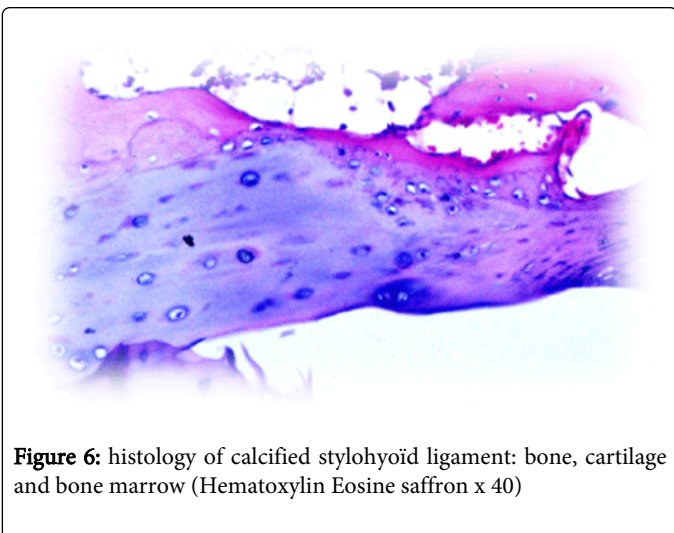


Figure 6: histology of calcified stylohyoid ligament: bone, cartilage and bone marrow (Hematoxylin Eosine saffron x 40)

## 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, cervicgia, 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 nervusvagus, which ties in with descriptions of reflex deaths [13]. The anatomical structures close to an elongation of the styloid process concern the nervusvagus [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 nervusvagus, 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

1. de la Grandmaison GL, Durigon M (2002) Sudden adult death: a medico-legal series of 77 cases between 1995 and 2000. *Med Sci Law* 42: 225-232.
2. de la Grandmaison GL (2006) Is there progress in the autopsy diagnosis of sudden unexpected death in adults? *Forensic SciInt* 156: 138-144.
3. Eagle WW (1937) Elongated styloid process. Report of Two Cases. *Arch Otolaryngol* 25: 584-587.
4. Eagle WW (1948) Elongated styloid process; further observations and a new syndrome. *Arch Otolaryngol* 47: 630-640.
5. Piagkou M, Anagnostopoulou S, Kouladouros K, Piagkos G (2009) Eagle's syndrome: a review of the literature. *ClinAnat* 22: 545-558.
6. Gossman JR Jr, Tarsitano JJ (1977) The styloid-stylohyoid syndrome. *J Oral Surg* 35: 555-560.
7. Keur JJ, Campbell JP, McCarthy JF, Ralph WJ (1986) The clinical significance of the elongated styloid process. *Oral Surg Oral Med Oral Pathol* 61: 399-404.
8. Vougiouklakis T (2006) Overview of the ossified stylohyoid ligament based in more than 1200 forensic autopsies. *J Clin Forensic Med* 13: 268-270.
9. Rao PP, Menezes RG, Naik R, Venugopal A, Nagesh KR, et al. (2010) Bilateral calcified stylohyoid ligament: an incidental autopsy finding with medicolegal significance. *Leg Med (Tokyo)* 12: 184-187.
10. Ruwanpura PR, Abeygunasekera AM, Tikiri GK (2008) Sudden unexpected death probably due to Eagle's syndrome: a case report. *Med Sci Law* 48: 350-353.
11. Doberentz E, Schyma C, Madea B (2012) Histological examination of the carotid bifurcation in case of violence against the neck. *Forensic SciInt* 216: 135-140.
12. Schrag , Vaucher P, Bollmann MD, Mangin P (2011) Death caused by cardioinhibitory reflex cardiac arrest--a systematic review of cases. *Forensic SciInt* 207: 77-83.
13. Amark P, Stödberg T, Wallstedt L (2007) Late onset bradyarrhythmia during vagus nerve stimulation. *Epilepsia* 48: 1023-1024.
14. Fusco DJ, Asteraki S, Spetzler RF (2012) Eagle's syndrome: embryology, anatomy, and clinical management. *Acta Neurochir (Wien)* 154: 1119-1126.
15. Lunetta P, Modell JH, Sajantila A (2004) What is the incidence and significance of "dry-lungs" in bodies found in water? *Am J Forensic Med Pathol* 25: 291-301.
16. Alboni P, Alboni M, Gianfranchi L (2011) Simultaneous occurrence of two independent vagal reflexes: a possible cause of vagal sudden death. *Heart* 97: 623-625.
17. Razak A, Short JL, Hussain SI (2014) Carotid Artery Dissection due to Elongated Styloid Process: A Self-Stubbing Phenomenon. *J Neuroimaging* 24: 298-301.
18. Farhat HI, Elhammady MS, Ziayee H, Aziz-Sultan MA, Heros RC (2009) Eagle syndrome as a cause of transient ischemic attacks. *J Neurosurg* 110: 90-93.
19. Tubbs RS, Loukas M, Dixon J, Cohen-Gadol AA (2010) Compression of the cervical internal carotid artery by the stylopharyngeus muscle: an anatomical study with potential clinical significance. *Laboratory investigation. J Neurosurg* 113: 881-884.
20. Schrag B, Mangin P, Vaucher P, Bollmann MD (2012) Death caused by cardioinhibitory reflex: what experts believe. *Am J Forensic Med Pathol* 33: 9-12.