Case report

Eagle syndrome: Is it a cause of death? – A case report

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Abstract:
Cardiovascular issues are mostly responsible for the sudden and unexpected deaths. However current case revolves around one abnormal anatomical finding of long styloid process-Eagle syndrome which can, though rare, be a cause of sudden and unexpected death.

Keywords: Eagle syndrome, Long Styloid Process, Sudden Death.

Introduction:
Around 10-15% of all deaths occur suddenly and unexpectedly (1). Sudden death is defined by the abrupt and unexpected death of a subject in good health within one hour (2). Depending upon the acuteness of death, either lethal cardiovascular diseases (approximately 50%) or chronic progressive diseases that remain largely clinically asymptomatic till death are primarily responsible. Diseases of respiratory tracts accounts for some sudden deaths (about 15%), as do gastrointestinal diseases (about 10%), diseases of the central nervous system (about 10%), and acute infection (1). However in the current case report we are discussing one very rare entity of Eagle syndrome which can be considered as one of the causes of sudden and unexpected death.

Case report:
A highly decomposed body with features of skeletonisation and natural separation of various joints was brought to the mortuary for the postmortem examination requesting the determination of exact cause of death. After careful examination we could not find any injury or fracture or any other finding to ascertain exact cause of death. Only abnormal anatomical finding we could see was abnormally long styloid processes of both sides. The skull was attached with extremely decomposed and thin layer of soft tissue. On each side the length of styloid process was 6 cm. (Image 1 and Image 2). The morphological features of the skull were of a male of age more than 50 years old age. Soft tissue and viscera kept for chemical analysis were negative for any poisonous substance. The cause of death could not be commented however possibility of death due to elongated styloid process in the current case could not be ruled out.
1. Image 1- Showing elongated left styloid process.

2. Image 2- Showing elongated right styloid process.

Discussion:
An abnormally long styloid process or stylohyoid ligament chain ossification producing cluster of symptoms gives rise to Eagle syndrome or stylohyoid syndrome (3). Eagle syndrome was first documented by Wart. W. Eagle in 1937 (4). The elongated styloid process and a calcified stylohyoid ligament manifesting its effect are seen only in 4% population (5). Some report a frequency of 1.4% in young subjects (6) while others indicate frequency of 30% for elderly (7). The styloid process may vary from 5 mm to 50 mm in length and stylohyoid ligament may ossify from its origin at the styloid process until its attachment with the hyoid bone (8). Only 4 to 10% of subjects afflicted presents with symptoms. The symptomatic group is characterised by preponderance of females and an age of over 50 (6, 7). Conversely an autopsy study relating to 1215 cases discovered frequency of 0.9% with male preponderance with only two cases with age not more than 30 years (9). In the current case the skull was of a male individual with age more than 50 years.

Eagle syndrome is subdivided into two clinical entities depending upon the different symptomatological characteristics noted subsequent to gathering 250 cases (10). The ‘standard syndrome’ is characterised by odynophagia, dysphagia, cervicalgia, and craniofacial pain. The ‘carotid syndrome’ entails faintness, feelings of intoxication, and even cerebrovascular accidents. In this area of anatomy the stylohyoid chain is in contact with juglar vein, carotid artery, vagus nerve and hypoglossal nerve (11). Rao PP et al has mentioned one case in which victim felt down as a consequence of fainting due to Eagle syndrome. The deceases had history of prior attacks of fainting type symptoms (12). In a case discussed by Ruwanpura et al a 39 year old lady showed history of faintness and headaches. Cause of death was attributed to Eagle syndrome supported by bilateral elongation of styloid processes measuring 3 cm and 3.7 cm along with partial calcification of stylohyoid ligament (13).

Clement R Barrios L reported a case in which 25 year old man died under circumstances suggestive of rotation and abrupt extension of neck, which according to author probably may have provoked a forced, exaggerated bearing down by vascular and lateral nerve elements of neck on the rigid structure of left stylohyoid chain which may have caused hypersensitivity of nervus vagus leading to reflex death (14).
In a case reported by Pradeep Kumar et al 55 year old female collapsed while sliding down from a height of 6ft. into a water body. Deceased had history of 2 episodes of collapse and frequent episodes of headaches prior to this fatal accident. She was labelled medically fit on prior occasions. During postmortem examination only positive and relevant finding was 4.75 cm long styloid process (15).

In present case no past history was available. Hence keeping in consideration all above case, only it could be said in present case that cause of death as Eagle syndrome could not be denied.

Changes in stretch and transmural pressure are detected by baroreceptors in the heart, carotid sinus, aortic arch, and other large vessels. Afferent impulses are transmitted by the carotid sinus and glossopharyngeal and vagus nerves to the nuclei tractus solitaries and the paramedian nucleus in the brain stem. Efferent limbs are carried through sympathetic and vagus nerves to the heart and blood vessels, controlling heart rate and vasomotor tone. In carotid sinus hypersensitivity, mechanical deformation of carotid sinus leads to exaggerated response with bradycardia or vasodilatation, resulting in hypotension, presyncope and syncope (16). Carotid sinus reflex death is due to vagus nerve impulses, which may cause the heart to stop beating resulting in cardiac arrest. The diagnosis of sudden death due to mechanical irritation of the carotid sinus is possible, when there are signs of acute cardiovascular failure, and other causes of death are out of question, along with ascertainments at the scene as well as observations by witnesses of the death process. It is also clear that a person may be completely symptom free or asymptomatic and still have carotid sinus hypersensitivity (17). A recent article refutes reflex death caused by fatal pressure in the neck without any histological lesion of the glomerulus (18). However several cases in literature describe vascular accidents caused by cases of Eagle syndrome involving extracranial carotid artery and not glomerulus (14).

In the current case in absence of,
1. Any positive previous history,
2. Complaints at the time of death and,
3. Obvious histological changes, one can only say that, in the current case cause of death is Eagle syndrome until otherwise proved.

**Conclusion:**
Eagle’s syndrome should be kept in mind as a cause in every single case of syncope. It is very difficult to label Eagle syndrome as cause of death in absence of positive past history and complaints. Postmortem diagnosis emphasises on histological changes, however they are not essential for labelling Eagle syndrome as cause of death.

**Ethical Clearance:** Not required.

**References:**