

CASE REPORT

Presentation of Eagle syndrome following radiation therapy to carcinoma of the larynx

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Abstract

Eagle syndrome is a rare clinical condition that is characterized by either an elongated styloid process or a calcified stylohyoid ligament. This report describes the case of a 35-year-old woman who presented with Eagle syndrome following the treatment of recurrent laryngeal carcinoma with ionizing radiation.

KEYWORDS

calcified stylohyoid ligament, Eagle syndrome, elongated styloid process, ossified stylohyoid ligament, radiation-induced fibrosis

1 | INTRODUCTION

In 1937, Watt Eagle first described the symptomology associated with an elongated styloid process and/or calcified stylohyoid ligament in two patients.¹ At that time, some of the associated symptoms were found to include dysphagia, throat pain similar to that of chronic pharyngitis, and foreign body sensation in the pharynx. Later studies found neuralgic pain of the throat, radiating pain to the ear, and odynophagia to be classically presenting symptoms as well.² Eagle also reported a different syndrome associated with an elongated styloid process, one in which the internal and/or external carotid arteries may be impinged upon.³ In more detrimental cases, elongated styloid processes have been shown to cause transient ischemic attack and carotid artery dissection.^{4,5}

Following a raised clinical suspicion for Eagle syndrome, plain X-ray may be used to confirm the diagnosis.

However, computed tomography (CT) remains the gold standard for diagnosis of Eagle syndrome due to its ability to more precisely examine soft tissue of the neck and more closely evaluate the proximity of an elongated styloid process to critical neuronal and vascular structures.^{6,7}

2 | CASE PRESENTATION

Our patient is a 35-year-old female who was initially referred to the otolaryngology clinic 6 years ago for the evaluation of hoarseness. Flexible endoscopy found a polypoid lesion of the left true vocal cord, which was determined to be moderately dysplastic squamous mucosa. Subsequent laryngoscopies displayed a progression to severe dysplasia and further to squamous cell carcinoma in situ with a small area of possible invasion. Our patient then underwent open left hemilaryngectomy with final pathology

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demonstrating T1a N0 invasive squamous cell carcinoma of the left true vocal cord. All margins were negative for invasive or in situ disease.

At 4 months post-op hemilaryngectomy, our patient was hoarse once again and underwent another laryngoscopy. Biopsy results demonstrated the recurrence of squamous cell carcinoma in situ of the left true vocal cord. The patient decided to forego another operation and proceeded with definitive radiotherapy. Image-guided radiation was delivered at a dose of 2 Gy per fraction for 33 fractions to a total of 66 Gy over the course of 51 days.

Five years post-definitive radiotherapy, our patient presented with left-sided anterior neck pain and odynophagia. The patient reported her neck pain to be constant and dull in nature with intermittent sharp pains occurring during lateral head movement in both directions. She reported the pain to have persisted over the course of approximately 3 months and to have used non-steroidal anti-inflammatory drugs and acetaminophen for symptomatic treatment. Regarding her odynophagia, the patient reported that it occurs intermittently with both solids and liquids, but the pain does not limit her oral intake. The patient's past medical and surgical history prior to hemilaryngectomy and subsequent radiotherapy was reviewed once again and was found to be non-contributory. Additionally, she denied any new medical or surgical history over the past 5 years that may have influenced her current presentation.

She was evaluated for Eagle syndrome. CT scan of the neck and soft tissue with contrast displayed elongation of the left styloid process to approximately 29.2 mm (Figure 1) without calcification of the stylohyoid ligament, correlating clinically with Eagle syndrome. The right styloid process was found to be equally elongated; however, the patient continued to deny any right-sided pain. CT findings further demonstrated that neither styloid process impinged upon critical neurovascular structures, especially those contained within the carotid sheath. Fibrotic soft tissue of the neck was noted on this CT scan bilaterally, but the extent of fibrosis could not be precisely quantified.

The CT scan at the time of presentation was compared with one that was performed 2 months prior to initial radiation therapy (Figure 2). There was no appreciable difference in left styloid process length prior to radiation therapy and at the time of presentation.

The patient was informed of the nature of her condition, emphasizing that there was no urgent need for intervention as neither of her elongated styloid processes were infiltrating critical structures. Knowing this information, she elected to continue treating her condition symptomatically with non-steroidal anti-inflammatory drugs and acetaminophen as she finds her neck pain and odynophagia



FIGURE 1 Sagittal CT scan of the neck and soft tissue with contrast. CT was performed at the time of symptom presentation, demonstrating elongation of the left styloid process

to be bothersome but not substantially impacting her activities of daily living.

3 | DISCUSSION

Radiation therapy to the head and neck has been found to cause a myriad of potential long-term side effects that include but are not limited to osteoradionecrosis, secondary cancer, and tissue fibrosis.⁸ Radiation-induced tissue fibrosis of the head and neck is of clinical interest in this patient's presentation of Eagle syndrome.

The role of ionizing radiation in the development of tissue fibrosis has been well-established and is known to cause an inflammatory response that ultimately leads to decreased tissue compliance.⁹ Specifically, radiation induces direct DNA damage and formation of reactive oxygen species that injure both normal and neoplastic tissues.¹⁰ Neutrophils are the first subset of inflammatory cells to reach an area of tissue damage, releasing multiple inflammatory cytokines such as interleukin-1, interleukin-6, and tumor necrosis factor-alpha that further enhance inflammation of the irradiated field.^{11,12} Monocytes then infiltrate the area of inflammation and differentiate into M1 and M2 macrophages, the latter of which secretes both platelet-derived growth factor



FIGURE 2 Sagittal CT scan of the neck and soft tissue with contrast. CT was performed 2 months prior to the patient's initial treatment with ionizing radiation, demonstrating elongation of the left styloid process

(PDGF) and transforming growth factor-beta (TGF- β).¹³⁻¹⁶ Release of PDGF causes substantial fibroblast migration to the field, while release of TGF- β leads to the differentiation of these fibroblasts into myofibroblasts.^{15,17} Importantly, myofibroblasts will secrete extracellular matrix proteins, including collagen, in response to TGF- β and will produce the characteristic thickened and stiffened tissue known as fibrosis.^{18,19}

We suggest that the long-term fibrotic changes associated with ionizing radiotherapy be considered as a cause of this patient's delayed presentation of Eagle syndrome. Fibrotic changes of the head and neck may have lowered our patient's threshold to present with symptoms associated with an elongated styloid process. Unfortunately, there is no universal method by which to objectively quantify the degree of head and neck fibrosis as criteria for assessing these fibrotic changes are still being developed.^{20,21}

4 | CONCLUSION

To the best of our knowledge, this is the sole report of a delayed presentation of Eagle syndrome that only presented following ionizing radiation to the neck. Radiation

therapy has been well-demonstrated to alter normal structural and functional anatomy, especially that of the head and neck. Prior history of ionizing radiation should not be ignored when evaluating a patient who presented with a delayed onset of symptoms associated with an elongated styloid process as their late presentation may be due to radiation-induced fibrosis.

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None.

CONFLICT OF INTEREST

The authors declare that there are no competing financial or personal interests that may have influenced the production of this case report.

AUTHOR CONTRIBUTIONS

Jake K. Cartwright wrote the original draft of this report in addition to conceptualizing the project and making subsequent revisions. Francisco G. Moreno contributed to project design, assisted in revision, and supervised the overall production of this report.

ETHICAL APPROVAL

East Tennessee State University IRB was consulted and confirmed that IRB approval was not required for this study.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

DATA AVAILABILITY STATEMENT

All data used in this study are available from the corresponding author upon reasonable request.

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