J Hear Sci, 2019; 9(4): 33–36
DOI: 10.17430/1003481

A RARE CASE OF LATE DIAGNOSIS OF ELONGATED STYLOID PROCESS SYNDROME

Contributions:
A Study design/planning
B Data collection/entry
C Data analysis/statistics
D Data interpretation
E Preparation of manuscript
F Literature analysis/search
G Funds collection

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Abstract

Background: Eagle's syndrome comes from the name of the American otolaryngologist – Watt Weems Eagle, who in 1937 was the first to distinguish a separate disease entity for symptoms associated with excessive elongation of the styloid process of the temporal bone.

Case report: A 42-year old patient was admitted to the Department of Otolaryngology, Laryngological Oncology, Audiology and Phoniatrics in November 2018, in a planned mode, with persistent throat pain radiating to the right ear, right side head pain, tongue numbness and hoarseness. Symptoms of variable severity maintained for about 9 years, which is why the patient underwent bilateral tonsillectomy in October 2016. Due to persistent hoarseness, the patient was also qualified for micro-direct laryngoscopy with the removal of vocal cord nodules, the procedure was performed in March 2017. Despite the surgical treatment, the pain persisted. On the basis of the medical history, clinical observation, after analyzing the results of the tests with particular consideration of the neck CT scan with 3D reconstruction, the patient was diagnosed with right-sided Eagle's syndrome. The patient was qualified for surgery under general endotracheal anesthesia. In November 2018 partial resection of the right elongated styloid process was performed using an intraoral approach. After wound healing the patient reported complete pain relief.

Conclusions: The elongated styloid process syndrome is most frequently a diagnosis of exclusion. Lack of proper diagnosis often delays significantly the implementation of proper management and thus exposes patients to long-term struggle with pain. If non-specific throat pain persists, the Eagle's syndrome should always be considered, which can be treated effectively.

Key words: elongated styloid process syndrome • late diagnosis • Eagle's syndrome

UN CASO RARO DE DIAGNÓSTICO TARDÍO DEL SÍNDROME DEL PROCESO ESTILOIDE ALARGADO Y OSIFICADO

Resumen

Introducción: El nombre de "Síndrome de Eagle" proviene del nombre de un otorrinolaringólogo estadounidense, Watt Weems Eagle, quien en 1937 fue el primero en distinguir una entidad de enfermedad separada para los síntomas asociados con el alargamiento excesivo de la apófisis estiloides del hueso temporal.

Informe del caso: Paciente de 42 años ingresada de manera programada en noviembre de 2018 debido a dolencias persistentes en la Clínica de Otorrinolaringología, Oncología Laringológica, Audiología y Foniatría. La paciente refiere dolor de garganta irradiado a la oreja derecha, dolor en la mitad derecha de la cabeza, sensación de entumecimiento de la raíz de la lengua y ronquera. Las dolencias de diversa gravedad persistieron durante aproximadamente 9 años. En octubre de 2016, la paciente se sometió a una extirpación bilateral de la amígdala palatina. Debido a la ronquera persistente, la paciente también posteriormente fue programada para una laringoscopia directa para exéresis de nódulos en las cuerdas vocales y dicho procedimiento se realizó en marzo de 2017. A pesar del tratamiento quirúrgico, el dolor persistió. En base a la historia clínica, la exploración clínica y después del análisis de los resultados de los procedimientos diagnósticos, con especial énfasis en la tomografía computarizada del cuello con reconstrucción 3D, la paciente fue diagnosticado de síndrome de Eagle derecho. Se programó a la paciente para cirugía bajo anestesia endotraqueal general. En noviembre de 2018, se realizó la eliminación parcial de la apófisis estiloides excesivamente alargada con vía de acceso intraoral. Después de la curación de la herida, el paciente informó de una resolución completa del dolor.

Conclusiones: Síndrome del proceso estiloide alargado y osificado suele ser un diagnóstico de exclusión. La falta de un diagnóstico adecuado a menudo retrasa significativamente la implementación del tratamiento adecuado y, por lo tanto, expone a los pacientes a una lucha a largo plazo con dolencias. Si persiste el dolor de garganta inespecífico, siempre hay que considerar el síndrome de Eagle, que puede tratarse eficazmente.

Palabras clave: síndrome del proceso estiloide alargado y osificado • diagnóstico tardío • síndrome de Eagle

РЕДКИЙ СЛУЧАЙ ПОЗДНЕЙ ДИАГНОСТИКИ ШИЛОПОДЪЯЗЫЧНОГО СИНДРОМА

Аннотация

Введение: Название Синдрома Eagle происходит от имени американского отоларинголога - Уотта Уимса Игла, который в 1937 году первым выделил отдельное заболевание на основании симптомов, связанных с чрезмерным удлинением шиловидного отростка височной кости.

Описание случая: В ноябре 2018 года 42-летняя пациентка поступила в Клинику отоларингологии, ларингологической онкологии, аудиологии и фониатрии в плановом режиме по причине постоянной боли в горле, иррадиирующей в правое ухо, боли в правой стороне головы, онемения основания языка и хрипоты. Симптомы различной степени тяжести сохранялись в течение примерно 9 лет, поэтому пациентке была проведена двусторонняя тонзиллэктомия в октябре 2016 года. Из-за постоянной хрипоты пациентка была также направлена на прямую микроларингоскопию с удалением узелков голосовых связок, операция была проведена в марте 2017 года. Несмотря на хирургическое лечение, боль сохранялась. На основании истории болезни, клинического наблюдения, после анализа результатов обследований, особенно компьютерной томографии шеи с трехмерной реконструкцией, пациентке был поставлен диагноз шилоподъязычного синдрома с правой стороны. Пациентка была направлена на операцию под общим эндотрахеальным наркозом. В ноябре 2018 г. была проведена частичная резекция правого удлиненного шиловидного отростка через рот. После заживления раны пациентка отметила полное устранение боли.

Выводы: Диагноз шилоподъязычного синдрома чаще всего ставится путем исключения. Отсутствие правильного диагноза часто значительно замедляет применение соответствующего лечения и, таким образом, приводит к длительному болевому синдрому у пациентов. Если неспецифическая боль в горле сохраняется, всегда следует принять во внимание синдром Eagle, который можно эффективно лечить.

Ключевые слова: шилоподъязычный синдром • поздняя диагностика • синдром Eagle

RZADKI PRZYPADEK PÓŹNO ROZPOZNANEGO ZESPOŁU WYDŁUŻONEGO WYROSTKA RYLCOWATEGO

Streszczenie

Wstęp: Nazwa „zespół Eagle'a" pochodzi od nazwiska amerykańskiego otolaryngologa – Watta Weemsa Eagle'a – który w 1937 roku jako pierwszy wyróżnił odrębną jednostkę chorobową dla objawów związanych z nadmiernym wydłużeniem wyrostka rylcowatego kości skroniowej.

Opis przypadku: Pacjentka l. 42 przyjęta w listopadzie 2018 roku do Kliniki Otolaryngologii, Onkologii Laryngologicznej, Audiologii i Foniatrii w trybie planowym z powodu uporczywych dolegliwości: bólu gardła promieniującego do ucha prawego, bólu prawej połowy głowy, uczucia drętwienia nasady języka oraz chrypki. Dolegliwości o zmiennym nasileniu utrzymywały się od ok. 9 lat. W październiku 2016 roku wykonano u pacjentki obustronne usunięcie migdałków podniebiennych. W związku z utrzymującą się chrypką pacjentkę zakwalifikowano także do mikrolaryngoskopii bezpośredniej z usunięciem guzków głosowych, zabieg wykonano w marcu 2017 roku. Pomimo zastosowanego leczenia chirurgicznego dolegliwości bólowe nadal się utrzymywały. Na podstawie: wywiadu, obserwacji klinicznej, po analizie wyników badań, ze szczególnym uwzględnieniem badania CT szyi z rekonstrukcją 3D, rozpoznano u pacjentki prawostronny zespół Eagle'a. Chorą zakwalifikowano do leczenia operacyjnego w znieczuleniu ogólnym dotchawiczym. W listopadzie 2018 roku wykonano częściowe usunięcie nadmiernie wydłużonego wyrostka rylcowatego prawego z dostępu wewnątrzustnego. Po wygojeniu się rany w gardle pacjentka zgłosiła całkowite ustąpienie dolegliwości bólowych.

Wnioski: Zespół wydłużonego wyrostka rylcowatego najczęściej jest rozpoznaniem z wykluczenia. Brak właściwego rozpoznania często znacznie opóźnia wdrożenie prawidłowego leczenia, a tym samym naraża pacjentów na długotrwałe zmaganie się z dolegliwościami. W przypadku utrzymywania się niespecyficznych dolegliwości bólowych gardła zawsze należy rozważyć wystąpienie zespołu Eagle'a, który można skutecznie leczyć.

Słowa kluczowe: zespół wydłużonego wyrostka rylcowatego • późne rozpoznanie • zespół Eagle'a

Introduction

The term Eagle's syndrome comes from the name of the American otolaryngologist Watt Weems Eagle, who in 1937 was the first to distinguish a separate disease entity for symptoms associated with excessive elongation of the styloid process of the temporal bone [1]. The average length of the styloid process in an adult human ranges from 2 to 3 cm; it is classified as elongated when its length exceeds 3 cm [2–4].

Three main classifications of the prolonged styloid process are distinguished. The first, introduced by Eagle, distinguishes between classic and carotid artery syndrome. The classic form is a complication after tonsillectomy when scar tissue shifts toward the elongated styloid process. It is manifested by chronic throat pain, dysphagia, feeling a foreign body lodged in the throat, articulation disorders, atypical facial neuralgia, and pain on opening the mouth widely. These ailments are caused by irritation of the cranial nerves: trigeminal, facial, glossopharyngeal, and vagus. In the carotid artery form there is mechanical compression of the carotid arteries and sympathetic fibers by the styloid process during rotation of the head. Patients may report severe unilateral headache, pain on turning the head, eye pain, visual disturbances, and in extreme cases fainting [1,5,6].

Another proposed classification of elongated styloid process is the Langlais classification where the assessment of structural abnormality of the styloid process is based on radiological examination. Here, three types are distinguished:

Type 1 (elongated) – elongation of the styloid process > 3 cm;

Type 2 (pseudoarticulated) – elongation of the styloid process > 3 cm and divided into two parts joined by a pseudo-articulation;

Type 3 (segmented) – a multi-segment styloid process consisting of two or more segments [7].

The third applied classification, O'Carroll's, is based on an assessment of the position of the styloid process tip in relation to the mandibular foramen and here three types can be distinguished:

Type A – the styloid process tip is located above the mandibular foramen;

Type B – the styloid process tip is located between the mandibular foramen and the mandibular angle;

Type C – the styloid process tip extends below the border of the mandibular angle [8].

The mechanism behind Eagle's syndrome is still poorly explained. There are several hypotheses explaining its causes, including hyperplasia or metaplasia in response to trauma, excessive ossification during development, and endocrine disorders in women during menopause [9,10].

The aim of the study was to present a rare case of the elongated styloid process syndrome and diagnostic difficulties leading to late diagnosis of Eagle's syndrome.

Case report

A 42-year old female was admitted, in a planned mode, to the Department of Otolaryngology, Laryngological Oncology, Audiology and Phoniatrics at the Medical University of Lodz in November 2018 with persistent throat pain radiating to the right ear, ipsilateral head pain, tongue numbness, and hoarseness. The patient did not report a previous history of trauma. Symptoms of variable severity had continued for about 9 years, which is why the patient underwent bilateral tonsillectomy in October 2016. Due to persistent hoarseness, the patient had also been previously qualified for micro-direct laryngoscopy with removal of vocal cord nodules, a procedure performed in March 2017. Despite the surgery, pain persisted. The patient described the pain as stabbing and not responding to painkillers. She underwent several consultations with different specialists. The patient underwent an MRI of the temporomandibular joints and a neck ultrasound, with no sign of abnormality. To aid diagnosis, a neck CT scan with 3D reconstruction was performed in September 2018 (Fig. 1). This revealed elongation of the styloid processes, especially on the right side: about 35 mm on the right side and 32 mm on the left.

On the basis of the medical history, clinical observation, and the tests and scans – particularly the neck CT with 3D reconstruction – the patient was diagnosed with right-sided Eagle's syndrome. The patient was qualified for surgical procedure under endotracheal anesthesia, and in November 2018 a partial resection of the excessively elongated

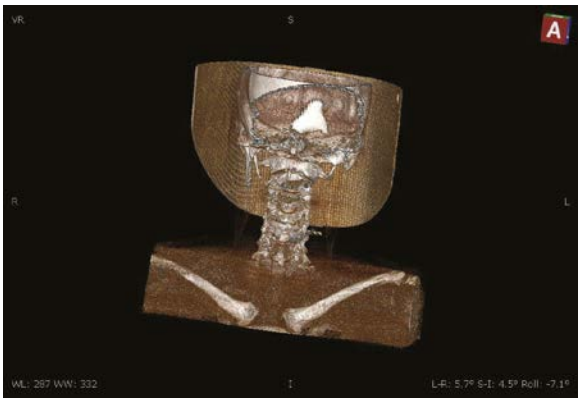

Figure 1. Computed tomography in the frontal plane with three-dimensional reconstruction (3D-CT) of the neck in the treated patient with visible elongated styloid process on the right side

right styloid process was performed using an intraoral approach. The surgery and postoperative period were uneventful. After recovery, the patient reported complete remission of pain and symptoms.

Discussion

Eagle's syndrome is a rare disease entity. It is a complex of symptoms that include recurrent throat pain, feeling of a foreign body lodged in the throat, headache, pain on neck rotation, otalgia, change in voice, and sensation of hypersalivation. Our patient complained of throat pain radiating to the right ear, headache, tongue numbness, and hoarseness.

Saccomanno et al. reported a rare case of a 60 year-old woman who presented severe unilateral trigeminal and glossopharyngeal neuralgia. The patient was subjected to conservative therapy for a few months which failed to relieve the symptoms. After several consultations with different physicians, a diagnosis was finally made based on a radiological examination (3D-CT). Surgical styloidectomy was performed with successful remission of symptoms [11].

On available reports, it has been estimated that the elongated styloid process occurs in 2–11.8% of the general population, 4% of which are symptomatic [12–15].

Bożyk et al. [4] assessed the length of the styloid process in 100 asymptomatic patients and the longest process was 72.2 mm.

In the available reports no correlation has been established between the frequency of the syndrome and gender, nor between the length of the styloid process and age [4].

Three-dimensional computed tomography reconstruction (3D-CT) is important for imaging elongated styloid process. Some authors also recommend cone beam computed tomography (CBCT), which also gives a three-dimensional image but uses much smaller dose of radiation. Conventional X-ray images present only a general picture of bone structures although can be informative in the initial diagnosis [4].

Treatment of elongated styloid process syndrome may be conservative or surgical. Conservative treatment is mainly used in the classic form of Eagle's syndrome. It includes injecting the tonsil lodge with anesthetics or glucocorticoids. Available reports also indicate the use of oral antiepileptic drugs or antidepressants as a non-surgical therapy.

Intraoral and extraoral approaches are used in surgical treatment. The unquestionable advantage of intraoral approach to the styloid process is shorter duration of the surgery and better cosmetic effects. Some authors draw attention to the higher risk of complications associated with limited visibility of the operating field compared to the extraoral approach [5,16]. Kiralj et al. have compared the two types of surgery. The authors reported cases of two male patients (49 and 34 years old) with elongated styloid process. Surgical involved unilateral right styloidectomy by the intraoral approach in the first case, and right styloidectomy by the extraoral approach in the second. Post-operative recovery was successful in both cases with no complications [17].

In the literature, transoral robotic styloidectomy has also been described as an alternative to traditional surgery [18].

Lack of proper diagnosis often delays proper treatment and exposes patients to a long struggle with their ailments.

The effectiveness of surgery in the treatment of elongated styloid process syndrome (in terms of pain relief and significant or complete remission of other symptoms) is about 80% [19].

Conclusions

1. An elongated styloid process syndrome is most frequently a diagnosis of exclusion.

2. Due to nonspecific symptomatology, it is often a diagnostic challenge for doctors of many specialties, but mainly otolaryngologists, neurologists, dentists, and family doctors.

3. Lack of proper diagnosis often significantly delays the introduction of proper treatment and exposes patients to long-term discomfort.

4. If non-specific throat pain persists, Eagle's syndrome should always be considered, a condition which can be treated effectively.

Written informed consent was obtained from the patient for the publication of this case report and accompanying image.

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