Case study

EAGLE SYNDROME: A CASE REPORT AND REVIEW OF THE LITERATURE

ABSTRACT

Eagle's syndrome or stylo-carotid syndrome, or elongated styloid process syndrome or American syndrome is a rare and controversial entity. It is characterized by a long styloid process causing irritation of the surrounding structures. We report a case of Eagle's syndrome of incidental discovery in a 53-year-old patient, with no particular history, who presented with intense left retro-mandibular pain evolving for 2 years. An angioscan of the supra-aortic trunks was ordered and showed two bilateral bony processes extending from the styloid process towards the lesser horn of the hyoid bone, indicating ossification of the stylohyoid ligament, which led to the diagnosis of Eagle syndrome.

Key words : Cervicalgia; Eagle syndrome; CT scan

INTRODUCTION

Eagle's syndrome is a rare radio-clinical entity characterized by a long styloid process or ossification of the stylohyoid ligament. It occurs when the stylohyoid complex conflicts with surrounding anatomical structures. It is a frequent cause of neck and craniofacial pain. This relatively frequent syndrome, which is not systematically investigated, is rarely diagnosed. Its diagnosis, according to Eagle [2], is clinical and confirmed by imaging means, especially CT scan [3-5], which plays a fundamental role in the surgical therapeutic management.

CASE REPORT

This is a 53-year-old female patient, with no particular pathological history, presenting with intense left retro-mandibular pain, increased by head rotation, associated with dysphagia, dysphonia with hypersalivation, all evolving for 2 years. An angioscan of the supra-aortic trunks was ordered, showing two bilateral bony processes extending from the styloid process towards the small horn of the hyoid bone, indicating ossification of the stylohyoid ligament (Figure 01).

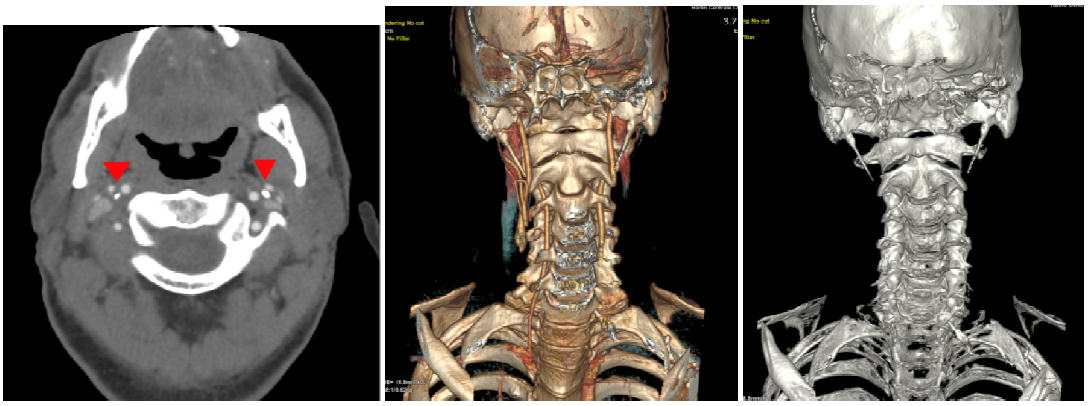

Figure 01: Axial scan section after PDC injection and in 3D reconstruction: showing elongation of the two styloid processes (red arrowhead) arriving at the carotid space of 34 mm in length on the right and 38 mm on the left.

DISCUSSION

Eagle's syndrome is a rare clinical entity. It was first described by Eagle in 1937 [1]. This syndrome is due to a long styloid process and/or calcification of the stylohyoid ligament and/or a long small horn of the hyoid bone [2]. The prevalence of long styloid apophyses measured in several cohorts of patients from radiological images varies around 4% but only 4% of them will develop a painful syndrome [1, 3]. Eagle syndrome occurs when the styloid process conflicts with adjacent anatomical structures: the carotid arteries, internal jugular vein, facial nerve, glossopharyngeal nerve, vagus nerve, and hypoglossal nerve [4, 5]. Most patients with Eagle syndrome are over 40 years old [4], with extremes of 20 and 80 years [6, 7]. In our series the age ranged from 46 to 60 years. The prevalence of gender in Eagle syndrome is highly controversial. The etiology of calcification or ossification of the stylohyoid ligament remains poorly understood, despite various theories without any scientific basis [9]. Indeed, most authors state that this syndrome usually follows one or more traumas or surgical procedures that took place near the styloid process (tonsil removal or dental surgery).The functional signs are variable, Eagle distinguished three groups, the first is the classic syndrome associating cervicalgia, otalgia and pharyngeal discomfort, the second characterized by pain along the external carotid artery and the third asymptomatic. Palpation of the tonsillar fossae allows the diagnosis to be suspected.

Standard radiological exploration confirms the diagnosis by showing the presence of the bony process that extends from the styloid process to the homolateral small horn of the hyoid bone. The superimposition of several bony structures and magnifications secondary to angulation are potential drawbacks of conventional radiography [11].With CT, these drawbacks are eliminated. In fact, the CT scan specifies the relationship of the calcified hyoid styloid ligament with the neighboring vascular and nerve structures [4]. Recently, 3D CT is a valuable tool in the diagnosis of Eagle's syndrome because of its ability to provide all information regarding the styloid process, including its length, direction and anatomical relationships [12].

Eagle syndrome can be treated by medical and surgical means. Non-surgical treatment consists of reassurance and pain relief with analgesics and injections of non-steroidal anti-inflammatory drugs and steroids [6, 8]. Some authors suggest the combination of xylocaine with corticosteroids [1]. The palpable tip of the styloid process, the tonsil fossa and the small horn of the hyoid bone are the injection points [1]. In the literature, this conservative treatment is effective in the short and medium term, but requires repetitive injections in the long term [10, 11]. Surgical treatment can be performed using one of two approaches: trans pharyngeal or extra buccal [10, 12]. There may be persistence of the symptomatology after surgical treatment, which leads us to ask questions about the effectiveness of the operative technique [10] and the presence of a possible associated but unrecognized pathology.

CONCLUSION

Eagle's syndrome is a relatively easy clinical diagnosis. Imaging means, in particular CT, allow not only an evaluation of the elongation of the styloid process but also the appreciation of its surrounding structures within the framework of a pre-therapeutic assessment. However, this syndrome remains unrecognized in many cases. It should therefore be considered in the presence of neck pain and vertigo without any observed cause.

REFERENCES

1. Eagle WW. Elongated styloid process. Arch Otolaryngol 1937;25: 584–7.
2. Colby CC, Del Gaudio JM. Stylohyoid complex syndrome: a new diagnostic classification. Arch Otolaryngol Head Neck Surg 2011;137:248-52.

3. Ait Ali Yahia D, Tanguy JY, Nedelcu C, Bouvier, Delorme B, Gourdier AL et al. Syndrome d'EAGLE ou syndrome de l'apophyse styloïde. SFR 2013. http://pe.sfrnet.org. (consulté le 14 Juin 2014)
4. Eagle WW. Elongated styloid process; further observations and a new syndrome. Arch Otolaryngol 1948; 47(5):630-40.
5. Raina D, Gothi R, Rajan S. Eagle syndrome. Indian J Radiol Imaging 2009;19(2):107-8.
6. Savranlar A, Uzun L, Uğur MB, Ozer T. Three-dimensional CT of Eagle's syndrome. Diagn Interv Radiol 2005;11(4):206-9.
7. Ramadan SU, Gokharman D, Tuncbilek I, Kacar M, Koşar P, Kosar U. Assessment of the stylohoid chain by 3D-CT. Surg Radiol Anat 2007; 29(7):583-8
8. Eagle syndrome: about three cases and review of the literature adigo a.m.y.1 , adjenou k.v. 1 , adambounou k. 1 , sonhaye l 1 , agoda-koussema l.k. 1 , djagnikpo o. 1 , boko e.n.2 , n'dakena k. 1 rev. cames sante vol.3, n° 1, juillet 2015
9. Valerio CS, Peyneau PD, de Sousa AC, Cardoso FO, de Oliveira DR, Taitson PF and al. Stylohyoid syndrome: surgical approach. J Craniofac Surg. 2012; 23: e 138-40.
10. Shivani J, Ashok B, Samrity P, Deepti V.P. Styloid-stylohyoid syndrome. Ann Maxillofac Surg 2012; 2(1): 66–69.
11. Colby C,Del Gaudio J. Styloid complex syndrome.Arch Otolaryngol Head Neck Surg 2011; 137(3): 248-252
12. Raina D, Gothi R, Rajan S. Eagle syndrome. Indian J Radiol Imaging 2009;19(2):107-8.