Open Journal of
Clinical & Medical
Case Reports

Volume 5 (2019)
Issue 14

ISSN: 2379-1039

Raeder syndrome and fibromuscular dysplasia without carotid artery dissection: A new association

Joao Brainer Clares de Andrade*; Jay P Mohr

***Joao Brainer Clares de Andrade**
Columbia University, Doris and Stanley Tananbaum Stroke Center, USA
Phone: +121-2305-8033; Email: joao.brainer@unifesp.br

Abstract

Background: Raeder Syndrome (RS) is described as unilateral ptosis and miosis and paratrigeminal syndrome without sweating abnormalities. Internal carotid artery dissection is the most reported underlying condition in patients with RS.

Case: A 49-years-old woman complains right-sided headaches featuring occasional droopy eyelid and blurred vision on the same side which have begun 6 years ago. The patient presented ptosis of the right eyelid with blurred vision and eye tearing and redness with cluster attacks. The patient had a diagnosis of Fibromuscular Dysplasia (FD) with classical features in bilateral extracranial internal carotid without dissection. Neuroimaging workup found no evidence of intracranial lesions, arterial dissection or cervical aneurysms.

Conclusion: A chronic distention of carotid sheath as seen in FD may damage the sympathetic fibers, can lead to Raeder Syndrome. To best of our knowledge, this is the first described association between RS and FD in a patient without carotid artery dissection.

Keywords

raeder syndrome; fibromuscular dysplasia; Headache attributed to cervical carotid or vertebral artery disorder; carotid disease

Abbreviations

RS: Raeder syndrome; FD: Fibromuscular dysplasia

Introduction

Raeder Syndrome (RS) is an uncommon condition described as unilateral ptosis and miosis and paratrigeminal syndrome, differentiating from Horner Syndrome in that ipsilateral sweating is preserved1. In addition, cranial nerve palsy may be described in patients with RS including other symptoms like ipsilateral eyelid edema, lacrimation and rhinorrhea [1,2].

Most reported cases of RS have been considered idiopathic [2], but pericarotid lesions have been pointed as a cause of RS [3]. Internal carotid dissection is the carotid-disease most associated with RS [2,4].

Raeder syndrome is commonly described as a benign and auto-limited condition [2]. From best of our knowledge Other carotid underlying conditions such as fibromuscular dysplasia without arterial dissection or carotid aneurysms have not been associated with RS at this time.

We aimed to describe the case of a patient with confirmed fibromuscular dysplasia with no evidence of internal carotid artery dissection with a long-term Raeder Syndrome.

Case Report

A 49-years-old woman with complaints of right-sided headaches featuring occasional droopy eyelid and blurred vision on the same side which have begun 6 years ago. The patient reported pain affecting the forehead and periorbital region and pounding in character (in clusters), lasting as long as several days with the same features. She denied any premonitory symptoms, transient somatosensitive, cognitive or auditory deficit or photophobia. The patient reported ptosis of the right eyelid with blurred vision and eye tearing and redness with cluster attacks as well. She saw a neuro-ophthalmologist - who found impaired adduction of her left eye. She denied any facial sweating abnormalities. Her family history is unremarkable. On exam, we saw a slight adductor bias with the left eye at rest head position and impaired adduction of the left eye. The trigeminal nerve response to sensory testing concluded that all 3 divisions were intact. Visual acuity, visual fields and pupils were normal.

The patient had a diagnosis of Fibromuscular Dysplasia (FD) with classical features in bilateral extracranial internal carotid, and left subclavian, left vertebral and right renal arteries corroborated with doppler and arteriography studies. Her last computerized tomography angiogram showed multisegmental areas of stenosis in the distal extracranial internal carotid bilaterally, and strongly suggestive for FD (Figure 1). At that time, there was no evidence of carotid artery dissection. A follow-up brain Magnetic Resonance Imaging performed in December 2018 came back negative for any abnormalities. The patient is under a well-tolerated pharmacologic treatment.

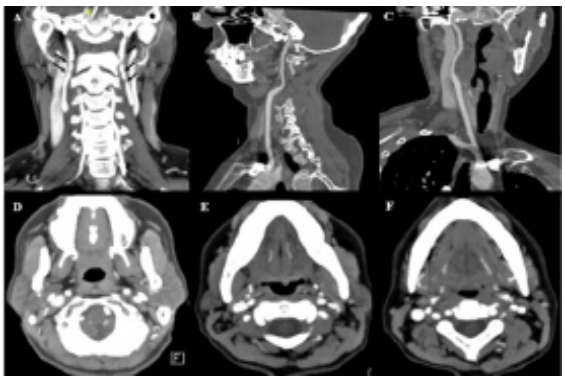

Figure 1: Neuroimaging workup with clear FD findings without evidence of carotid artery dissection. **Legends:** Coronal (A) and sagittal (B, C) cervical CT-angiogram in maximal intensity projection (MIP) demonstrating carotids with multiple saccular dilations and stenosing webs (black arrows). This finding is compatible with the classical “strings of beads” sign. There is no evidence of carotid artery dissection (D-F).

Discussion

The first description of Raeder Syndrome symptoms dates back to the beginning of the last century [1,2]. Raeder was originally described as a relationship between paralysis of the oculopupillary sympathetic nerve and pain in the trigeminal nerve distribution, implicating the paratrigeminal area of the middle fossa of the cranium" [2] - and the first case reported was in an 18-year-old laborer with pain in the left eye which spread to the left temple, cheek and left superior jaw, including ptosis of the upper lid and miosis on the left eye.

The symptoms may begin in a few days or weeks following a possible injury, and they may disappear within some months or rarely persist for several years [1-3]. The symptoms may also disappear or improve after the treatment for the underlying condition [1,2].

The Raeder Syndrome physiopathology is based on the way of sympathetic fibers are projected. Sympathetic fibers emerge between the C6 and T4 spinal segment and go upward superior cervical ganglion on the sheath of internal carotids – the fibers that control sweating on the ipsilateral side of the face project from external carotids sheath to the skin [1,3] , and it means that RS is a consequence to injuries of internal carotids. Part of these fibers is directly linked to the trigeminovascular system. This system is activated via antidromic conduction by some neuropeptides delivered on terminals of the system, leading to an aseptic neurogenic inflammation – which actives trigeminal projections to the thalamus [5].

Our patient has long-term facial pain with intermittent autonomic and neurological deficits. Her neurological exam, considering her complains, is compatible with autonomic dysfunction from sympathetic fibers projecting on the internal carotid, leading to a trigeminal facial pain with the reported features. Also, she has a history of refractory to some profilatic drugs to migraine – which emphasizes a persistent condition working as a trigger to pain.

Many cases of RS have been attributed to head or cervical trauma, viral infection, arteriosclerosis disease, aortitis, maxillary sinusitis, chronic otitis and lobar pneumonia [1,2,6,7], including neurosyphilis8 – which a perivasculitis may explain the phenomenon [3,8].

Other carotid diseases (aneurysm, pseudoaneurysm and inflammatory disease) have been associated with RS1. The authors suggest that in addition to the mechanical compression, the inflammatory status may lead to disruption of sympathetic fibers along the carotid sheath [1,9].

Our patient performed an extensive neuroimaging work up – which found no evidence of arterial dissection or important arterial enlargements such as aneurysms. Thus, we think that FD may lead to a perivascular disease – which may explain the high frequency of symptoms for a long time for our patient, despite the presence of that two cited conditions [5].

Fibromuscular dysplasia is a nonatherosclerotic or inflammatory vascular disease that may lead to arterial stenosis (maybe a consequence of the intimal fibroplasia), aneurysm or dissection [10,11]. This

condition is most prevalent in young and females; the average age of diagnosis is 5.9 (13.4) years, ranging from 5 to 86 years old [12]. Sharing a similar physiopathology with RS and being considered as the same condition for a long time, Horner Syndrome has also a rare association with FD in patients without arterial dissection [9,10].

Options to manage and treat RS associated with FD are scarce in the literature. Other conditions like arterial dissection or severe arterial stenosis have been proposed to treat with stents or angioplasty. As for our patient, we are concerned that an angioplasty could make a possible intermittent syndrome to persist indefinitely.

Conclusion

This is the first association between RS and FD described in a patient with no prior history of internal carotid artery dissection. Persistent RS is another uncommon point in our case, at this time. All these findings, regarding published cases about fibromuscular dysplasia and Raeder Syndrome or Horner Syndrome, emphasize that a chronic distention of carotid sheath as seen in FD may lead to damage of sympathetic fibers – which may lead to autonomic dysfunction and recurrent paratrigeminal syndrome.

Clinical Implications

- Raeder Syndrome is a non-described condition in patients with Fibromuscular Dysplasia without evident carotid artery dissection until now.
- Internal carotid enlargements may lead to progressive dysfunction of sympathetic fibers along its sheath
- Raeder Syndrome may be considered a no time-limited condition in association to Fibromuscular Dysplasia with carotid arter involvement

Sources of Funding

Dr. Andrade's visiting scholarship at Columbia University, New York City, is sponsored by the Capes Foundation, Ministry of Education, Brazil

Ethical Approval

All ethical principles have been followed. Written informed consent was obtained from the patient for the publication of this case report

References

1. Healy JF, Zyroff J, Rosenkrantz H. Raeder syndrome associated with lesions of the internal carotid artery. Radiology. 1981; 141:101–104.

2. Solomon S. Raeder Syndrome. Archives of Neurology. 2001; 58.

3. Vijayan N, Watson C. Pericarotid Syndrome. Headache: The Journal of Head and Face Pain. 1978; 18: 244–254.

4. Hart RG, Easton JD. Dissections of Cervical and Cerebral Arteries. Neurologic Clinics. 1983;1: 155–182.

5. Tatsui CE, Prevedello DM-S, Koerbel A, Cordeiro JG, Ditzel LFDS, Araujo JC. Raeders syndrome after embolization of a giant intracavernous carotid artery aneurysm: pathophysiological considerations. Arquivos de Neuro-Psiquiatria. 2005; 63: 676–680

6. Solomon S, Lustig J. Benign Raeders Syndrome is Probably a Manifestation of Carotid Artery Disease. Cephalalgia. 2001; 21: 1–11.

7. Raeder's syndrome. Report of two cases. Int Angiol [Internet]. 1995Dec;14(4):415–417.

8. Watanabe K, Tanahashi N, Nara M. Neurosyphilis presenting with raeders syndrome. Annals of Neurology. 1989; 25: 418.

9. Mcinnis CP, Haynor DR, Francis CE. Horner syndrome in fibromuscular dysplasia without carotid dissection. Canadian Journal of Ophthalmology. 2016; 51.

10. Mettinger KL, Ericson K. Fibromuscular dysplasia and the brain: observations on angiographic, clinical and genetic characteristics. Stroke. 1982; 13: 46–52.

11. Olin JW, Gornik HL, Bacharach M, et al. Fibromuscular dysplasia: state of the science and critical unanswered questions. Circulation. 2014; 129: 1048-1078.

12. Olin JW, Froehlich J, Gu X, et al. The United States Registry for Fibromuscular Dysplasia: results in the first 447 patients. Circulation. 2012; 125: 3182-3190.

Manuscript Information: Received: August 17, 2019; Accepted: September 24, 2019; Published: September 30, 2019

Authors Information: Joao Brainer Clares de Andrade[1,2*]; Jay P Mohr[1]
[1]Columbia University, Doris and Stanley Tananbaum Stroke Center, USA
[2]Universidade Federal de Sao Paulo, Sao Paulo, Brazil

Citation: Andrade JBC, Mohr JP. Raeder syndrome and fibromuscular dysplasia without carotid artery dissection: A new association. Open J Clin Med Case Rep. 2019; 1586.