

A Rare Cause of Horner Syndrome: Arteria Lusoria

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Received date: July 25, 2017; Accepted date: September 19, 2017; Published date: September 24, 2017

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Abstract

Horner syndrome is a combination of clinical signs, classically of ipsilateral ptosis, pupillary miosis and facial anhidrosis, secondary to the interruption of the oculosympathetic pathway. The causes include tumour infiltration, compression by a lesion such as an aneurysm, iatrogenic causes and traumatic injuries. This paper presents a case of Horner Syndrome due to a rare cause, a congenital anomaly of the aortic arch, arteria lusoria. A 37 year old, female patient referred to our department with a 4 weeks history of ptosis in the right eye. Her medical history was unremarkable. On examination, there was ptosis at her right eyelid and right pupil was myotic. After the tests and examinations, her clinical problem was decided as a well-known condition named "Horner syndrome". Our patient diagnosed as Horner syndrome secondary to aberrant right subclavian artery (arteria lusoria) which is the most common congenital anomaly of the aortic arch. Horner syndrome may be a result of common arterial anomaly like arteria lusoria. Although, there is still not an exact guideline for the diagnostic approach of Horner syndrome and to follow the diagnostic steps according to the patients' condition would be helpful to find the definite underlying pathology of Horner syndrome.

Keywords: Horner syndrome; Arterial Anomaly; Arteria Lusoria

Introduction

Horner syndrome is a combination of clinical signs, classically of ipsilateral ptosis, pupillary miosis and facial anhidrosis, secondary to the interruption of the oculosympathetic pathway [1]. The causes include tumour infiltration, compression by a lesion such as an aneurysm, iatrogenic causes, and traumatic injuries [2]. This paper presents a case of Horner Syndrome due to a rare cause, a congenital anomaly of the aortic arch, arteria lusoria.

Case Report

A 37 years old, female patient referred to our department with a 4 weeks history of ptosis in the right eye. She did not report fluctuation or diplopia during the day. Her medical history was unremarkable. On examination there was ptosis at her right eyelid and right pupil was myotic. In normal room lighting, the pupils measured 4 mm at left eye and 2 mm at right eye. In the dark, the size of the pupils changed to 5 mm at left eye and 2 mm right eye.

No inverse ptosis (elevation of the lower lid) was present, but there was approximately 2 mm of right upper lid ptosis. We instilled 1% apraclonidine in each eye prior to any other pharmacologic agent that day. The patient was evaluated 1 hour later, and in normal room lighting, the left pupil remained 5 mm.

The right pupil increased from 2 mm to 4 mm, resulting in a reversal of the anisocoria. Other neurological examination findings

were normal, but there were multiple palpable cervical lymph nodes on the systematic examination of the patient.

With these examination findings the patient was evaluated as partial Horner's syndrome. According to etiological approach for first order lesions of the Horner syndrome (central; posterior hypothalamus, brainstem and cervical spinal lesions effecting intermediolateral grey substance at the levels of C8-T2) [1,3].

Magnetic resonance imaging (MRI) of the brain, MR-angiography and MR-venography of the brain and cervical spinal MRI were performed and all were unremarkable. After excluding a central pathology, the patient was examined by an otorhinolaryngologist for the cervical lymphadenopathies. Hematologic evaluations to exclude lymphoproliferative diseases were also performed.

Otorhinolaryngologists recommended that using a two-week antibiotic therapy with the diagnosis of reactive cervical lymphadenopathy, however her complaints did not resolve after that treatment. At the same time, the patient was evaluated for the second order lesions (C8-T2 ventral nerve roots, apex of the lung, mediastinum and cervical sympathetic chain [1,3]) and the third order lesions (Superior cervical ganglion at the C2-3 carotid artery, Cavernous sinus, orbit [1,3]) of the Horner's syndrome.

Her neck MRI and orbital MRI were normal, however thorax computerized tomography (CT) and thorax CT angiography (TCA) showed an aberrant right subclavian artery (Arteria lusoria) (Figures 1 and 2).

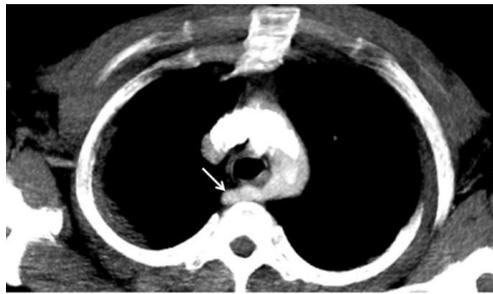


Figure 1: Images from thorax CT angiography. Showing an aberrant right subclavian artery (Arteria lusoria) (arrow).



Figure 2: Images from thorax CT angiography. Showing an aberrant right retroesophageal subclavian artery in the reconstructed images (arrow).

Discussion

Our patient presented with common clinical signs of Horner syndrome and finally diagnosed as Horner syndrome secondary to aberrant right subclavian artery (arteria lusoria) which is the most common congenital anomaly of the aortic arch [4]. Horner syndrome is one of the presenting symptoms of aberrant right subclavian artery, however majority of the patients with this anomaly never develop clinical signs and symptoms.

In the diagnostic steps of Horner syndrome, it is really difficult to decide the right emergent diagnostic test. Frequently, the patients have no associated symptoms to determine or localize the underlying pathology and it is difficult to choose the most important diagnostic approach. The diagnosis of Horner's syndrome begins with a detailed clinical history with use of adjunctive pharmacological testing and diagnostic imaging. In this case the results of the apraclonidine test are compatible with the diagnosis. But, as the apraclonidine test can similarly confirm the diagnosis but is unable to localize a site for targeted imaging [5,6], we continued the diagnostic investigations with further imaging tests.

The anatomical pathway of the oculosympathetic chain is too long, so we decided to start the diagnostic tests to exclude central and emergent pathologies, however these tests were unremarkable. The underlying pathology of the Horner syndrome is reported to be apparent in over 80% of patients at the time of the first neuro-ophthalmic consultation, based on history or clinical localization of the lesion [1]. But this case does not have an apparent etiology. It is reported that urgent imaging is not always required for patients with Horner syndrome with no localizing signs like pain, trauma, malignancy [1,7]. As there were multiple palpable cervical lymph nodes which were diagnosed as reactive cervical lymphadenopathy by the otorhinolaryngologists, the patient was put on a two-week antibiotic therapy, but her symptoms did not recover. Davagnanam et al. recommended performing a CT angiogram within the six weeks of submission in the absence of localization such as our patient [1]. In according to recommended diagnostic steps, we performed a TCA and it showed an aberrant right subclavian artery. As we could not differentiate the first, second and third order neuron pathologies with pharmacological tests in this patient, the definite diagnosis could be found by the help of CT angiogram of the arcus aorta.

Aberrant right subclavian artery is known as an arch variant and some of the patients with this variant anomaly may present with aneurysmal degeneration and adjacent aortic pathology [8]. Our case does not have any pathology like aneurysmal degeneration and adjacent aortic pathology. She was consulted with vascular surgeons, but she did not accept any surgical procedure. It is reported that arteria lusoria is more common in females like in our patient and dysphagia is the major symptom of it in adults [9,10]. This condition is called "dysphagia lusoria" [9] which usually present in the fourth or fifth decades [11] and esophageal compression of a rigid trachea seems to be its cause. The close relationship between the aberrant right subclavian artery, oesophagus, trachea and oculosympathetic pathway is responsible for various presentations seen among these patients. Compression of oesophagus and trachea may cause dysphagia, dyspnea or pneumonia [12] and compression of oculosympathetic pathway may cause Horner Syndrome. Such symptoms worsen with age when compression becomes more pronounced as oesophageal rigidity and atherosclerotic hardening of arteries increase. In our patient, Horner syndrome was the only clinical sign and the atherosclerotic changes in the walls of the arteries and increased compression of oculosympathetic pathway may be the underlying pathology.

Conclusion

In conclusion, it is important to remember that Horner syndrome may be a result of a common arterial anomaly like arteria lusoria. Although, there is still not an exact guideline for the diagnostic approach of Horner syndrome and the diagnostic steps according to the patients' condition would be helpful to find the definite underlying pathology of Horner syndrome.

Conflict of Interest

The manuscript is not under simultaneous consideration by any other journal. The manuscript has not been previously published. All co-authors have made a significant contribution to the manuscript. The study complies with ethical standards. The authors declare that they have no financial interests. None of the authors have any conflict of interest.

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