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Pourfour du Petit syndrome caused by traumatic pseudo-aneurysm of the internal carotid artery

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Abstract. Pourfour du Petit syndrome, or reverse Horner syndrome, is described as an overactive sympathetic nervous system, being characterized by mydriasis, eyelid retraction, and hyperhidrosis. We described a case of Pourfour du Petit syndrome after cervical injury by gunshot, with a little review about this rare syndrome. Angiography revealed dissection and formation of pseudo aneurysm of the left carotid artery. We believe that this lesion caused hyper-stimulation of the left cervical sympathetic chain, resulting in reverse Horner syndrome or Pourfour du Petit syndrome. There was reversal of symptoms spontaneously after 3 wk.

Keywords: Horner syndrome, autonomic nervous system, Pourfour du Petit syndrome, carotid artery dissection

1. Introduction

Horner syndrome is, characterized by ptosis, miosis, and hemifacial anhidrosis, caused by inhibitory lesion in the ipsilateral cervical sympathetic chain [1]. Pourfour du Petit (PDP) syndrome, or reverse Horner syndrome is, described as an overactive sympathetic nervous system being, characterized by mydriasis, eyelid retraction, and hyperhidrosis [2].

The present case report describes a case of a patient; victim of gunshot wound in the cervical region that showed the reverse Horner syndrome. We also carry out a brief review of the literature on the subject.

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2. Case report

A male patient, 16 yr, was victim of a gunshot wound in the cervical region; bullet entry was in the left submandibular region, transfixing the cervical spine. The patient was found unconscious by the rescue team with signs of respiratory failure, and underwent endotracheal intubation. He underwent a head computed tomography, which showed ischemic area in the left cerebral hemisphere. The patient underwent intensive care under sedation, until clinical and hemodynamic stabilization. Diagnostic complementary assessment with brain angiography demonstrated evidence of dissection of the left internal carotid artery in its cervical segment with pseudo aneurysm formation (Fig. 2).

After discontinuation of sedation, a neurological examination revealed anisocoria (left larger than

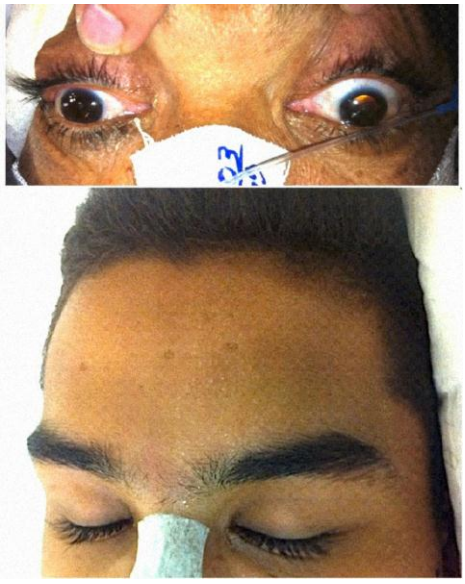

Fig. 1. Anisocoria with left larger than right (up), and sweating on left side (down). (Colours are visible in the online version of the article; http://dx.doi.org/ 10.3233/JPN-130615)

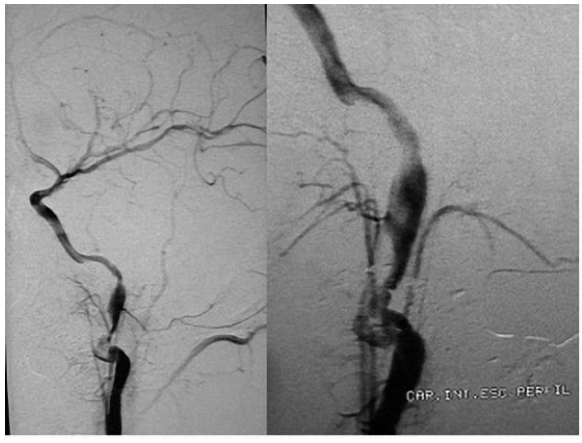

Fig. 2. Arteriography demonstrated evidence of dissection of the left internal carotid artery with pseudo-aneurysm formation in its cervical segment.

right), as showed in Fig. 1. When in a dark room, unlike Horner syndrome, the difference between pupil sizes decreased. When one eye was illuminated (left or right), both pupils contracted. The left side of the face showed sweating (Fig. 1) and mild blush. The mydriasis associated with hyperhidrosis and ipsilateral eyelid retraction characterized PDP syndrome. The patient was still aphasic due to ischemia in the left carotid territory, he was underwent endovascular treatment of carotid dissection. Patient evolution showed reversal of the syndrome, and he was isochoric 3 weeks after the event with improvement of general status.

3. Discussion

François Pourfour du Petit provided the first description of symptoms of what would later known as PDP syndrome in the eighteenth century, who attributed its alterations to the hyper stimulation of the sympathetic fibers, in opposition to Horner syndrome, in which there is inhibition of the sympathetic pathway [1–3]. He also performed experiments in dogs [4], reproducing clinical signs of the syndrome.

In classical Horner syndrome, there is ptosis, miosis and anhidrosis in the affected side. In the PDP syndrome, there is eyelid retraction, mydriasis and hyperhidrosis, as shown in this case. For this reason, PDP syndrome is also known as reverse Horner syndrome [2]. In both syndromes, dilated pupils are reactive to light, however, in the dark room, the difference between the sizes of pupils in PDP syndrome decreases, while in Horner syndrome, this difference increases.

Preganglionic sympathetic neurons that innervate the head arise from the first two thoracic levels (T1 and T2) pass through the lower and middle cervical ganglia to synapse in the ganglion superior. Postganglionic fibers of superior cervical ganglion cells innervate blood vessels and smooth muscle, as well as the sweat, lachrymal and salivary glands of the head [5]. The reverse Horner syndrome, or PDP syndrome, has been reported as a phenomenon that can be caused by irritation of the sympathetic nerves on the affected side and may result in Horner syndrome [6]; however, the pathophysiology is not elucidated [2,5,6]. Other syndromes caused by injury to the cervical sympathetic chain are also described as harlequin syndrome, which is expressed with facial flushing and sweating, explained as being secondary to an excessive compensatory response by an intact sympathetic pathway ipsilateral, when the contra lateral side has a sympathetic deficit [7].

There are no epidemiological data about PDP syndrome, with information due only to cases reported since its description. PDP syndrome has been reported in association with carotid dissection [8], intracranial aneurysms [3], cervical contusion and brachial plexus injuries [9], post-traumatic syringomyelia, severe

brain injury, aortic malformations and thyroid carcinoma [1]. In this present case, we believe that the sympathetic hyperstimulation occurred as a result of the cervical lesions caused by the projectile and its fragments. This injury led to internal carotid artery dissection and pseudo aneurysm formation, which may have stretched the fibers of the cervical sympathetic chain causing ipsilateral stimulus to these fibers. PDP evolution is variable: the clinical characteristics can be maintained indefinitely or show remission of some of its signs in a few mo [1], or may also result in Horner syndrome [6,8]. In the presented case, PDP symptoms were gone at three weeks spontaneously, even before the endovascular treatment of arterial dissection.

In conclusion, PDP syndrome is a rare syndrome, sometimes confused with Horner syndrome, however a neurologically concise examination is enough to differentiate them. In Horner syndrome, the ipsilateral pupil to cervical injury is miotic, while in the PDP syndrome is mydriatic.

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