Primary Cough Headache Associated with Jugular Insufficiency: Report of Two Cases

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Abstract

Background: Insufficiency of the internal jugular valve is a scarcely found etiology in the literature for the cough headache, which can be associated with other vascular disorders. The objective of this study is to report, through clinical and imaging findings, the rarely described association between cough headache and jugular insufficiency. Cases: Two male patients, ages 79 and 70-years respectively, were evaluated with episodes of headache associated with the Valsalva Maneuver. The first patient presented with a holocranial headache and two episodes of associated syncope after cough, while the other patient reported headaches after use of an abdominal belt, as a preventive measure to contain abdominal hernia. In both cases, the insufficiency was proved upon jugular reflux on Doppler sonography. Conclusion: The two case reports help to broaden the discussion on the possible association between primary cough headache and insufficiency of the internal jugular vein system, although the correlation between those two pathologies is still being debated.

Key Words: Primary cough headache; Valsalva Maneuver; Jugular Vein; Venous Insufficiency; Doppler Ultrasound (Source: MeSH-NLM).

Introduction

Primary cough headache (PCH) has a prevalence in the general population of 1%, more frequent in males, usually self-limited and with prolonged remissions.1 The pain has a sudden onset (<10 seconds), reaches a plateau, and lasts for seconds to minutes until it completely disappears after 120 minutes. The cough headache syndrome may be associated with intracranial structural changes: Chiari type I malformation, vascular disorders of the carotid and vertebral arteries; tumors of the middle and posterior fossa; basilar impression; platybasia; reversible cerebral vasospasm syndrome, among others. Cases with undefined etiology are classified as probable PCH. This type of primary headache usually does not present with associated symptoms such as nausea, vomiting, photophobia, phonophobia or autonomic manifestations.2

A functional jugular valve prevents retrograde flow into the internal jugular vein.3 In opposition, vein reflux has been associated with events of transient global amnesia,3 physical exertion headache,4 idiopathic intracranial hypertension associated with transverse sinus stenosis,3 cough headache when related to uremia and deep vein thrombosis,5 and a single report of cough headache without other associated pathologies.6

The association of insufficiency of the internal jugular valve with PCH is rarely described. In this work, we report two cases, present their clinical aspects, and discuss the proposed mechanisms of this association. Jugular insufficiency was considered as the presence of venous reflux flow lasting >0.88 seconds in the Doppler sonography as done previously in the literature.7 This case report was approved by the Ethics Committee of the Federal University of Paraná General Hospital (registration 4.523.357; Certificate of Presentation of Ethical Appreciation number: 42357721.8.0000.0096) and written informed consent was obtained from both patients prior to data collection.

Case #1

A 79-year-old man of Portuguese heritage, Body Mass Index (BMI) of 25.9 kg/m², reported headaches with initial onset eight months ago associated exclusively with Valsalva Maneuver (VM) such as when coughing or sneezing. He denied previous history of headaches. The headache manifested as high intensity pain in
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the right temporoparietal region with immediate holocranial distribution, of sudden onset, reaching peak intensity in 10 seconds, maintaining the same characteristics for 60 seconds, and easing within 10 minutes. No associated symptoms were described, and no abnormalities were detected on neurological examination. Brain Magnetic Resonance Imaging (MRI) showed moderate signs of supratentorial microangiopathy and small foci of hemosiderin deposition by micro-hemorrhages. Brain Magnetic Resonance Angiography (MRA) with both arterial and venous phases only showed signs of atheromatosis with irregularities in the vertebro-basilar system. The venous phase did not show any type of alterations suggestive of venous congestion such as arteriovenous malformations. Doppler sonography of the jugulars showed insufficiency of the Right Internal Jugular Vein (RIJV) with a reflux of 0.9 seconds during VM (Figure 1A). The left vein had a reflux of 0.7 seconds. Being clinically classified as having PCH, the patient was treated with 10 mg of propranolol and 1 capsule of horse chestnut (Aesculus hippocastanum), every 12 hours, with an improvement of 70% in pain intensity and frequency. Indomethacin, the first choice for PCH, could not be used because the patient had gastritis with consequent intolerance to the drug.

**Figure 1a.** Blood flow in the right internal jugular vein (RIJV) at rest (above) and in the Valsalva Maneuver (below). **Figure 1b.** Blood flow in the right internal jugular vein (RIJV) at rest (above) and in the Valsalva Maneuver (below).

**Legend:** Figure 1a - The reflux confirms the insufficiency of the RIJV (below) in case #1. Figure 1b - In the second patient (case #2), blood reflux is noticed after the Maneuver, which reinforces the insufficiency of the RIJV (below).

**Case #2**

A 70-year-old man of Germanic heritage, BMI of 29.1 kg/m², reported onset of headaches one year ago after starting the use of a belt to control abdominal hernia post-cholecystectomy. He presented with two distinct headache pain patterns. The first was continuous, located on the right occipital region; of moderate intensity, radiating to the cervical region and aggravated by head extension, cough, and digital compression on the ipsilateral major occipital nerve. It was diagnosed as major occipital nerve neuralgia and controlled with an anesthetic block of the respective nerve. The second pain pattern was explosive, localized to the right temporoparietal region, with subsequent holocranial distribution, triggered by cough, reaching maximum intensity in 8 seconds and lasting up to 3 minutes, with improvement and pain resolution in 15 minutes. No associated symptoms were described and neurological examination was within normal range. It was diagnosed as PCH. Previously, he only had episodes of migraine without aura, becoming asymptomatic after 50 years of age. Brain MRI and MRA (arterial and venous phases) only showed diffuse cerebral atrophy and discrete atherosclerosis, respectively. The venous phase of the MRA also did not show any type of alterations suggestive of venous congestion. Doppler sonography demonstrated the presence of insufficiency of the RIJV with a reflux of 1.1 seconds with re-production of pain symptoms (similar to the PCH) during the VM (Figure 1B). The left vein had a reflux of 0.5 seconds. The patient was treated with the same therapeutic combination used in the first patient, with an improvement of 40% in pain. Indomethacin could not be used because the patient, coincidentally, also had gastritis. However, once the belt was removed, the patient became totally asymptomatic from the second headache pain pattern.

**Figure 2.** Internal Jugular Valve in Function.

**Legend:** A. System in normal function during diastole and systole without triggering nociceptive trigeminal fibers. B - Jugular valve insufficiency during systole and without Valsalva Maneuver. C - Jugular valve insufficiency during diastole, associated with Valsalva Maneuver and pre-valvular ingurgitation subsequent to nociceptive trigeminal trigger.

**Discussion**

We describe two cases of PCH, and specifically in the second case, the patient re-presented with his headache symptoms during the VM on the Doppler sonography that identified the presence of an associated right jugular insufficiency. The diagnosis of this primary headache entity is established according to the criteria of the International Classification of Headache Disorders 3rd edition.
(ICHD-3), which are cited as follows: A - minimum of two headache episodes satisfying criteria B to D; B - caused by and taking place only in association with coughing, straining and/or other VM; C - sudden start; D - duration between one second and two hours; E - not better explained by another ICHD-3 diagnosis. All the above mentioned factors were observed in our patients.

According to some authors, the associated jugular venous insufficiency could be a factor related to the pathogenesis of PCH, although some also have suggested that this entity could be considered a secondary headache when venous insufficiency is demonstrated. The classification of these patients is complex, since it is not possible to directly relate the headache with jugular insufficiency, which is found in healthy individuals, with a prevalence of about 20-40% and increasing with age and obesity.

Headache associated with the reflux of the internal jugular vein during diastole can be justified by increased intracranial pressure; cerebral edema; venous edema, and stimulation of trigemino-cervical nociceptors located on the surface of the venous system. Doeppl et al. found that the retrograde flow of the jugular vein may play an important role in the pathophysiology of primary exertional headache, showing a 70% prevalence of jugular failure in symptomatic patients versus 20% in the asymptomatic control group.

In a letter, Knappertz first hypothesized that patients with cough headaches had incompetence and/or a lack of functionality of the internal jugular valve, accentuated during VM, due to the increase in intrathoracic and intra-abdominal pressures. The retrograde venous flow, due to insufficiency, produces an intravenous swelling with dilation of the plexus in the pre-vascular region, a site innervated by nociceptors from the trigeminal system, commencing with pain and resulting in trigeminal stimulation (Figure 2). This effect is not only in the pre-vascular region, but also having fluctuations in pressure causes an equidistant change from this location, as shown by a study that evaluated the pressure inside the sigmoid sinus at rest (160 mmH2O) and during the VM (425 mmH2O) in a patient with PCH and internal jugular valve failure.

The VM is a controlled method for a better understanding of how the cough acts in the human body, both producing similar physiological effects in increasing the venous pressure within the chest and abdomen. This system is richly formed by veins that Anastomose without the presence of a suitable valve system, mainly around the spinal canal. The transmission of the dilation of this venous system over the dura mater produces a collapse (pressure) on the neuraxis, increasing the pressure of the cerebrospinal fluid (lumbar pressure > cisternal pressure), which produces an upward wave in the lumbar cisternal direction, followed by a pressure balance and a subsequent reduction of lumbar pressure by inverting this wave from descending (cisternal - lumbar) and returning to the normal resting state. This is one of the reasons why VM can result in an increase or appearance of headache due to increased intracranial pressure.

After VM, the blood withdrawn in the venous system returns to the heart, producing an immediate cardiac output, thereby leading to peripheral vasoconstriction with a transient increase in blood pressure and heart rate, and a subsequent compensatory bradycardia. The more intense or persistent these autonomic changes, the more it can alter the control of the intracerebral neurovascular unit, reducing pain thresholds. Due to the physiology involved in VM and its compensatory responses and the consequences of a jugular insufficiency, the occurrence of headache in both reported cases shows a possible causal association between these factors. However, evidence demonstrating this direct etiological correlation is lacking in the literature. Therefore, our article brings a favorable occurrence to the association of a headache caused by VM and jugular venous insufficiency.

According to the current literature, Indomethacin is the recommended therapy for patients with PCH. Both patients had clinical contraindications for the use of this medication, being prescribed propranolol in these cases. Horse chestnut is occasionally used in the treatment of chronic venous insufficiency, although not directly associated with benefits in jugular vein insufficiency.

The report of these two cases is not able to prove the efficacy of the therapy with horse chestnut on the patients’ symptoms, and therefore, more studies are required on the drug therapy of this specific condition. On the other hand, the subjective outcome was satisfactory according to both patients and the removal of the abdominal belt resulted in a complete resolution of the headache in the second case, strongly suggesting the correct diagnosis and treatment of this patient. That said, the clinical outcome of these cases can be summarized in good prognosis and control of the PCH in the long term, even if an ideal treatment is still unclear.

Conclusion

Two patients are described with symptoms consistent with PCH, who also had jugular venous insufficiencies verified in Doppler sonography, which may or may not favor the occurrence of this primary headache. Although this association was clinically observed, it is not yet possible to determine a correlation between these two pathologies, hence reinforcing the necessity of further clinical studies.

References


