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New daily persistent headache – rare primary or secondary phenomena caused by carotid-cavernous fistula? A case report.

Case Report

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Abstract: Carotid cavernous fistulas (CCF) are dural arteriovenous fistulas which include pathological communications between the arterial system and the venous cavernous sinus situated at the wall of the cavernous sinus. It can be demonstrated by wide range clinical presentations. The presented case shows a very modest clinical presentation of CCF which was present over a few months only as new daily persistent headache without initial positive findings on performed computer tomography of the head, but rapidly worsened over a period of few days.

Keywords: *Carotid cavernous fistula • New daily persistent headache*

1. Introduction

Carotid cavernous fistulas (CCF) are rare pathological conditions characterized by abnormal arterial venous communication within the cavernous sinus. According to the currently used angiographic criteria there are direct and indirect types of CCF. The direct type of CCF means a direct flow from the internal carotid artery (ICA) to the CCF, while the indirect type of CCF means an indirect flow from the dural branches of the internal or external carotid system. Based on flow level through these fistulas, direct CCF is also known as high flow and indirect CCF is revealed as low flow malformation [1–3]. The clinical manifestations of CCF are caused by its location and size, blood drainage and collateral blood network. Generally, CCF is presented as bulbomotor palsy, diplopias, chemosis, and loss of visus, trigeminal nerve injuries and epistaxis [4].

Beside many reports designed to present a wild variety of CCF clinical presentations, and also relatively well defined clinical algorithms for CCF diagnosis, it seems there should be alert the earliest diagnostic tools for identifying CCF because CCF carry a high probability of intracranial hemorrhage or neurologic deterioration [5].

Thus, what authors highlight, as the most important in the next presented case, is initially poor clinical presentation of CCF, with silent neurological expression (mimics new daily persistent headache), without abnormalities on noninvasive imaging studies, but blown up neurological progression within few days.

2. Case report

A twenty eight year old woman has been presented with 4 months history of bilateral headache, predominantly localized deep in the left eye. Headache was daily and

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unremitting from within 3 days of its onset, of a non-pulsating quality, mild to moderate intensity (5-6/10 using visual assessment scale), without worsening with routine physical activity. In rare attacks headache was associated with mild nausea. Headache was partially reduced after non specific analgotherapy. Patient has not had prior headache` history. There were not ophthalmoplegia, proptosis and chemosis. Neurological status was revealed as physiological.

After initial headache onset computed tomography (CT) of the head was performed without observed positive findings.

According to International Classification of Headache Disorders–ICHD-3 criteria [6] actual headache was defined as a new daily persistent headache (NDPH), a self-limiting subform that typically resolves within several months without specific therapy.

Whereby headache was persistent without obvious improvements, enhancing in pain intensity, with severe nausea and vomiting, 4 months later patients was hospitalized and CT angiography was performed. Lateral and anteroposterior left carotid angiogram showed high flow (CCF) with brisk shunting from the left internal carotid artery into the botha cavernous sinuses and drainage into dilated superior ophtalmic vains and facial veins; and reflux into cortical veins (a poor prognostic factor). There was no supply of the distal internal carotid artery and intracranial arteries (Figure 1 and 2). Anteroposterior right carotid angiogram showed contralateral supply of the left anterior cerebral artery (ACA) and medial cerebral artery (MCA). There was not supply of the CCF from the right ICA (Figure 3).

Three days after admission to the Clinic, fully left side ophtalmoplegia with diplopia, left side ptosis, chemosis and hypestesis in ophthalmic area have occurred in neurological examination. The patient was scheduled for endovascular therapy. Stent assisted coil embolization with complete occlusion of the CCF was performed with complete recovery.

3. Discussion

The usual clinical presentation of a direct high-flow CCF appears as a consequence of high-pressure arterial blood transmitted directly into the cavernous sinus and ophthalmic veins, leading to venous hypertension [7], which can include a various presentations such as proptosis, chemosis, conjunctival injection, cranial nerve pareses, and visual deficits, bleeding (from mouth, nose, or ears), and cerebral complications (intracranial hemorrhage, increased intracranial pressure, and steal phenomena) [8].

However, CCF presentations are often vague in onset and then progress when the fistula arises [9]. There are reports which suggest that headache associated with the above mentioned signs can be a usual presentation of CCF, in up to 40-50% cases [10,11], but rare are those which point out headache as the only clinical symptom of CCF. These, CCF associated headaches, in small number of cases, can be presented as mimicking primary headaches [12], but in our patients it was a rare clinical phenotype of primary headache, NDPH. This type of headache is one of the most common and often

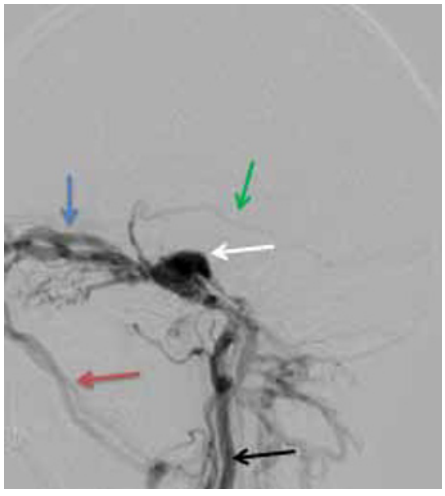

Figure 1

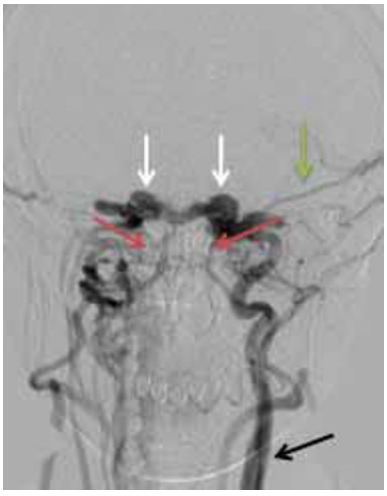

Figure 2

Figure 1. and 2. Lateral and anteroposterior left carotid angiogram showing high flow (CCF) with brisk shunting from the left internal carotid artery (black arrow) into the botha cavernous sinuses (white arrows) and drainage into dilated superior ophtalmic vains (blue arrow) and facial veins (red errows); and reflux into cortical veins (green errows)(a poor prognostic factor). There is no supply of the distal internal carotid artery and intracranial arteries.

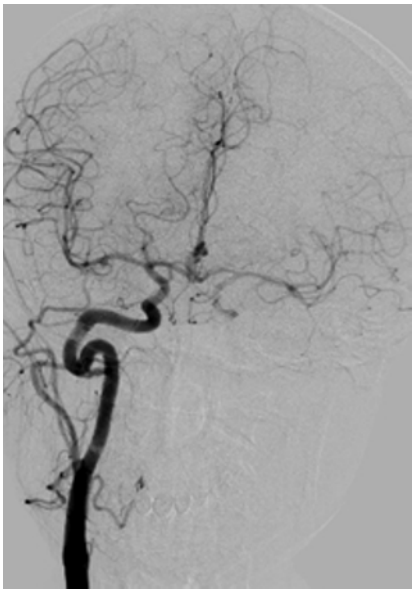

Figure 3. Anteroposterior right carotid angiogram showing contralateral supply of the left anterior cerebral artery (ACA) and medial cerebral artery (MCA). There is not supply of the CCF from the right ICA.

difficult headache disorders for diagnosis and therapy. Guidelines suggest making diagnosis of NDPH after the exclusion of many possible causes of secondary headaches by history, radiological examination and testing [13,14].

Typical findings of noninvasive cerebral imaging with CT scanning and/or MRI can be suggestive of CCF [13]. On the other side, it must be borne in the mind that the absence of abnormalities on noninvasive imaging studies (as was in initial CT scanning in here presented case) does not exclude the diagnosis of CCF [13]. Thus, cerebral angiography presents the gold standard imaging modality in the diagnosis of CCF, which is useful in identifying the localization of fistula, evaluating venous drainage and assessing collateral circulation [12,13]. While low risk CCF with mild symptomatology can be managed conservatively, higher risk fistulas deserve the most aggressive approach in order to eradicate the fistula. It includes surgical managements, radiosurgery and endovascular interventions such as detachable balloon occlusion, coil and material embolization (performed in our patients), covered stent graft placement, parent artery occlusion and transvenous or transarterial embolization [14].

Although all diagnostic criteria for NDPH can be fulfilled [6], in the conditions when the cause of a headache is not surely excluded as well as without positive CT/MRI findings, diagnosis of headache, suggestive for NDPH, must be taken guardedly, examining headache in the direction of its secondary etiology, i.e. CCF. The shown case is significant because the CCF in our patient had a very modest clinical presentation which was present over a few months, and which was, based on the clinical characteristics and absence in initial noninvasive imaging study's findings, regarded as NDPH, but which was rapidly worsened over a period of three days.

Therefore, clinicians should reconsider making the correct diagnosis of CCF performing cerebral angiography as obliged, in all NDPH patients suspected on CCF, in order to provide adequate therapy and preventing sudden clinical progression, intracranial hemorrhage and neurologic deterioration.

4. Conflict of interest

No conflict of interest exists for any of authors listed in the article.

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