Diagnosis and treatment of headache probably attributed to cerebral venous sinus thrombosis

Abstract
Cerebral venous sinus thrombosis (CVST) is a rare condition whose most common and sometimes only symptom is headache. Alas, diagnosis and treatment of CVST is often delayed or overlooked because of its high clinical variability. Using guidelines advises in detecting warning signs or symptoms of secondary headaches might ease the diagnosis of CVST. The article presents the case of a woman who is in treatment for chronic migraine and assessed for secondary headache in a multidisciplinary outpatient headache program. Alert symptoms like sudden worsening headache presentation, along with anamnestic cues, prompted neuroimaging that detected left transverse sinus thrombosis whose onset was difficult to date.

Keywords: Primary Headache; Secondary Headache; Chronic Migraine; Cerebral Venous Sinus Thrombosis

Diagnosi e terapia di una cefalea probabilmente derivante da trombosi dei seni venosi cerebrali

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INTRODUCTION
Cerebral venous sinus thrombosis (CVST), a rare condition whose incidence is 3-4 cases per 1 million people (75% of cases are women) and up to 7 cases to 1 million among children, is the presence of a blood clot in the dural venous sinuses draining deoxygenated blood from the brain [1]. The most common and sometimes only symptom of CVST is headache (80-90% of patients), while visual symptoms, stroke symptoms, seizures and depressed level of consciousness can be present as well [1,2]. The diagnosis may be suspected on the basis of symptoms, for example the combination of headache, signs of raised intracranial pressure, and focal neurological abnormalities [1,2]. The diagnosis is confirmed by neuroimaging showing thrombus obstructing the venous sinuses [1,2,3]. CVST risk factors include: thrombophilia, chronic inflammatory diseases, blood disorders, use of estrogen-containing contraceptives, meningitis, injury to the venous sinuses, medical procedures in the head and neck area, sickle cell anemia, dehydration, homocystinuria [1]. Therapy options are anticoagulants and rarely thrombolysis along with therapy for the underlying cause. In case of complication by raised intracranial

Why we describe this case
This case provides an example of a situation where is needed to understand that a secondary headache might be in place in a patient with a history of treated chronic migraine, even when acute neuroimaging markers are not detectable anymore. A multidisciplinary headache assessment and management in a hospital outpatient program proved effective in this case.

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pressure, surgical placement of a shunt can be performed. Symptoms like headache are also treated [4,5,6]. Alas, the diagnosis and treatment of CVST is often delayed or overlooked because of its high clinical variability [7]. The rate of missed diagnoses and treatment can be reduced using a diagnostic and therapeutic path able to detect symptoms and signs of the CVST presentations and orientate the most appropriate care.

**CASE PRESENTATION**

We present the case of M. L., a 35-year-old Italian woman living in Calabria. Following a neurological consult in January 2014, she was referred to the Headache Center at the Pugliese-Ciaccio Hospital in Catanzaro to continue receiving the Botulinum Toxin type A (BTXA) treatment for chronic migraine previously received in a hospital located in another region of Italy. The referral aimed to eliminate the need for long travels to receive therapy.

The patient came to the Headache Center in Catanzaro in March 2014. At a first medical examination, where a headache diary was given to the patient, emerged that the patient was not previously assessed for secondary migraine before or after receiving chronic primary headache treatment. Therefore, Mrs. L. was admitted to a chronic migraine outpatient program aimed to perform a complete headache assessment and subsequent therapeutic prescriptions. The program, that is multidisciplinary (it includes neurological examination, psychological evaluation, disability evaluation, pharmacological assessment, neuroimaging exams, on a need-basis specialist consults and pharmacological and non-pharmacological prescriptions) is designed to be adaptable to the patient presentation, needs, and preferences. As a first step in the program (T1), a medical history assessment, a neurological examination and a psychological evaluation were scheduled and performed in March 2014.

Medical history wise, blood hypertension was present in the familiar medical history in both maternal and paternal sides. Mrs. L. was married, had no kids, hold a university degree in economics and worked as an accountant; she had no history of any substance use or abuse; her personal medical history included: common exanthems in childhood; hyperomocisteinemia treated with daily intake of calcium mefolinate; endometriosisis treated with two surgical procedures in 2005; ovarian dysfunction treated with daily oral contraceptives; Mrs. L. was in treatment since 2009 for chronic migraine with BTXA and used diclofenac pills to manage acute episodes of migraine since 1998. Botulinum therapy had initially reduced the frequency of headache days from 25 days/month to 6 days/month.

General physical examination did not show anything to report. Neurological examination showed cranial hypoesthesia; Mrs L. reported suffering of prolonged and repeated fronto-temporal pulsating headache attacks lasting up to one week whose pain intensity increased over the days. Attack treatment with non-steroidal anti-inflammatory drugs was not effective. Reportedly, gait instability and recurrent vomiting had developed. Pain intensity classified by Numeric Rating Scale (NRS) was 8 out of 10. Headache days had risen to a value of 15 per month.

Psychological evaluation consisted in a clinical interview and the administration of self-report questionnaires to measure anxiety and depression level and to detect pathological anxiety or mood state, the Zung Anxiety Self Report Scale (Zung-Anx) and the Zung Depression Self Report Scale (Zung-Dep). Each of those tests consists of...
20 affirmations. The subject had to express her degree of agreement on a 4-point scale (from 1 = never to 4 = always). Psychological assessment showed no occurrence of pathological anxiety or mood state, nor any pathological personality trait (Zung-Anx = 33; Zung-Dep = 37). Migraine Disability Assessment (MIDAS) was administered in order to evaluate quality of life impact of the headache in the patient. MIDAS score (108) showed high level of disability experienced by the patient in work, social, and familiar life.

A pharmacological use assessment remarked continuous use of diclofenac tablets (150 mg) as attack treatment since the chronic migraine onset but excluded medications overuse.

Following medical and psychological evaluations, considering the presence of a secondary headache risk factor (usage of oral contraceptives) along with a possible alert symptom (sudden change and refractory worsening of the headache presentation), neuroimaging examinations were prescribed and performed, in order to investigate secondary forms of headache: nuclear Magnetic Resonance Imaging (MRI) and venous Magnetic Resonance Angiography (vMRA).

The MRI showed altered signal around the pineal gland without Fluid-Attenuated Inversion Recovery (FLAIR) signal suppression. The signal was judged possibly indicating a pineal cyst. Also, it showed alteration of the signal of the maxillary sinuses and turbinates of possibly inflammatory nature. The vMRA showed absence of the signal within the left transverse sinus and hypoplasia of sigmoid sinus and guls of the ipsilateral jugular (Figure 1).

Considering the vMRA result, a hematological consult to perform a genetic trombophilic investigation was requested and a prothrombin mutation (G20210A) was detected. As a diagnostic conclusion, a secondary headache, headache probably attributed to consequences of cerebral venous thrombosis along with primary chronic migraine, was diagnosed in March 2014 [5]. At the same time, the prescribed therapy was therefore adjusted in light of the recent diagnostic change: thrombolysis and/or oral anticoagulant were not prescribed to the patient because it was not possible to temporally date the CVST onset; In facts, signal hyper-intensity, acute venous thrombosis marker, was not evident in the MRI T1 sequence. Therefore, the vMRA finding was judged as a probably old dated thrombosis outcome. After a consultation with the local hospital Hematology and Thrombosis Center, costs-benefits ratio wise, the anticoagulant therapy was considered in-appropriate, since it was a long but undefined time ago dated thrombosis outcome. Topiramate (25 mg/day) was prescribed as a prophylactic therapy, while eletriptan (40 mg) was used as a once a need treatment. Moreover, the patient continued receiving BTXA (200 units per session) treatment every three months, being the next injection scheduled in April 2014. A follow-up program every three months after each injection session was planned. In June 2014, at the first follow-up examination (T2), the prolonged and pain increasing headache
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When to suspect a secondary headache?

Certain warning signs or symptoms may suggest a secondary headache, prompting additional examination (blood tests, brain scans, lumbar puncture) and/or referral to a specialist. These include:

- neurological symptoms during or between headaches;
- positive neurological examination;
- new or unusual type of headache;
- sudden or worst headache of one’s life;
- headaches unresponsive to treatment and/or steadily worsening;
- older age;
- immunocompromise;
- fever.

In the presented case, a sudden and refractory change in headache presentation, neurological signs along with a continuous contraceptive usage in medical history as a risk factor, prompted magnetic resonance and venous magnetic resonance angiography as additional examinations: neuroimaging detected cerebral venous sinus thrombosis as probably the underlying cause of a secondary headache. Nonetheless, there are no visible previous acute neuroimaging markers for a thrombosis that has probably happened a significant time before the examination.
REFERENCES


