Case Report

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An Unusual Presentation of Cerebral Vein Thrombosis: A 40-Year-Old Woman with Acute Intractable Cervical Pain and Lateral Neck Bending

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Received: 22 January 2018 **Revised:** 4 February 2018 **Accepted:** 02 March 2018 **Published:** 15 March 2018

ARTICLE INFO

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Keywords:
Venous thrombosis;
Neck pain;
Oral contraceptives;
Cranial sinuses

ABSTRACT

Cerebral venous thrombosis (CVT) is an uncommon but fatal type of stroke with a wide spectrum of signs and symptoms. We report a 40-year-old woman who presented with acute intractable neck pain and lateral cervical bending. Investigations with neuroimaging revealed evidence of thrombosis in her right transverse and sigmoid sinuses. The precipitating factors for the thrombosis were consumption of oral contraceptive pills during the month of Ramadan, and dehydration due to fasting. The patient immediately was treated with the therapeutic dose of anticoagulant. We concluded that in patients with severe cervical pain, especially those who are predisposed to thrombotic complications, CVT should be considered in the differential diagnosis.

Citation: Kalantar SS, Amirzargar N, Ranji-Burachaloo S. **An Unusual Presentation of Cerebral Vein Thrombosis: A 40-Year-Old Woman with Acute Intractable Cervical Pain and Lateral Neck Bending**. Case Rep Clin Pract 2018; 3(1): 30-4.

Introduction

Cerebral venous thrombosis (CVT) is an infrequent type of stroke, in which thrombosis of the dural sinuses or veins result in cerebral infarction, hemorrhage, or both. On one hand, CVT must be considered in the differential diagnosis for patients with a broad range of clinical presentation. On the other hand, to avoid progression of thrombosis and significant morbidity or mortality, early diagnosis and initiation of anticoagulation are obligatory (1, 2).

We present a case with a history of consumption of oral contraceptive pills (OCPs) who was admitted for severe right cervical pain, and lateral neck bending;

investigation with neuroimaging revealed evidence of thrombosis in right transverse and sigmoid sinuses.

Case Report

A 40-year-old woman was referred to our neurology clinic, suffering from severe right side cervical pain, and lateral neck bending started one week before, one day after the end of the Ramadan. She had taken high-dose (HD) OCPs continuously for 3 years during the month of Ramadan for religious purposes, due to its regulatory effect on menstruation period. The patient had visited several physicians during the week she experienced her current symptoms, and had received multiple analgesics and muscle relaxants, but her pain did not subside. The patient had an otherwise clear medical history.

On physical examination, she seemed to have an ill appearance, but her mental status appeared to be normal. Optic discs were sharp, eye movements were normal, and no meningismus signs were detected. Muscle forces, sensory examination, and cerebellar tests showed no abnormalities. She appeared to have severe tenderness at the right side of her neck, especially at the upper two third of the sternocleidomastoid muscle. No cervical lymphadenopathy, palpable mass, skin redness, or warmness existed. She seemed to bend her neck to the left side in order to decrease her pain. This view resembled torticollis; but no muscle spasm was detectable at any side. Although cervical soft or hard tissue pathologies were the first diagnosis speculated by the physicians, the absence of any response to analgesics and muscle relaxants, normal cervical plain X-ray, and more importantly, use of OCP put CVT in the upper part of the differential diagnosis list.

At first, the patient was admitted to the emergency department; then a brain computed tomography (CT) scan, D dimer level, and other routine biochemistry tests were requested. Excessive hydration with normal saline was administered whilst awaiting the results of the tests. The D-dimer test was positive (1990 ng/ml, with normal laboratory reference of 0-500). Suspected hyperdensity at right transverse sinus without hemorrhagic lesion was found in brain CT scan (Figure 1).

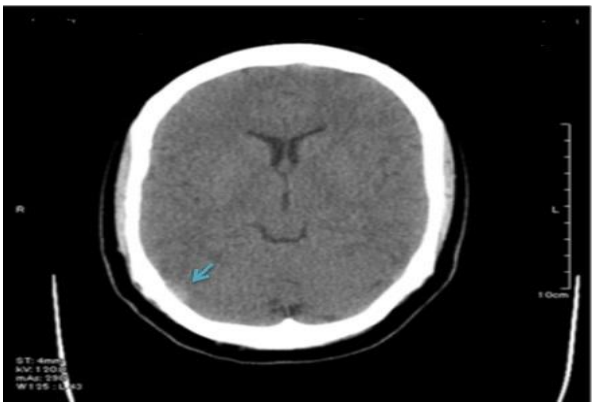

Figure 1. Axial brain computed tomography (CT) scan without contrast
Note the suspicious hyperdensity at the right lateral sinus. Although it can be considered as a Streak artifact, absence of similar changes in the other side, and repetition of hyperdensities in serial sequences in the route of lateral sinus, increases the suspicion of cerebral venous thrombosis (CVT).

Anticoagulant treatment with low molecular weight heparin (LMWH) at the therapeutic dose (enoxaparin 60 mg every 12 hours) was started. In brain magnetic resonance imaging (MRI) with and without gadolinium, and magnetic resonance venography (MRV) existence of a clot in the right transverse and sigmoid sinuses was confirmed (Figure 2).

The next day after receiving two doses of anticoagulant and hydration, the patient reported that her cervical pain had decreased by fifty percent.

Discussion

CVT is less frequent than arterial thrombosis; but if not diagnosed and treated early, may cause a sequel, or may be fatal. According to International Study on Cerebral Vein and Dural Sinus Thrombosis (ISCVT), albeit early recognition and treatment of CVT, the mortality during the acute phase is 5.6% (1).

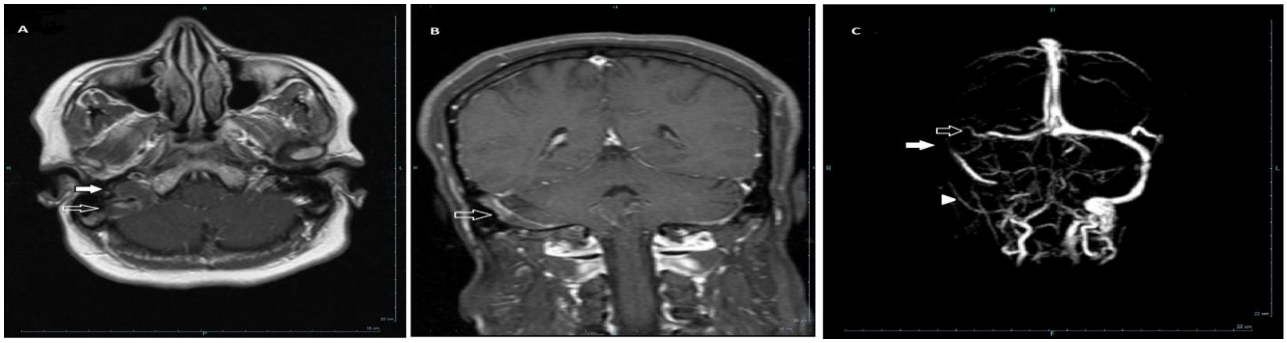

Figure 2. Thrombosis of the right transverse sinus (white arrow) and the right sigmoid sinus (black arrow) in T1-weighted image with contrast enhancement in axial (A) and coronal planes of brain magnetic resonance imaging (MRI) (B). Magnetic resonance venography (MRV) does not depict the right transvers and sigmoid sinuses and proximal part of internal jugular vein (arrow head) (C).

Early diagnosis and appropriate treatment are indispensable, as CVT may cause considerable complications including venous infarction, pulmonary embolism, subarachnoid hemorrhage, epilepsy, and dural arteriovenous fistula (2).

CVT is a diagnostic challenge to physicians because of the wide variety of symptoms and signs which range from altered consciousness, focal neurological deficits, and seizures to symptoms of raised intracranial pressure like headache, vomiting, and blurred vision. However, rare symptoms like thunderclap headaches, unilateral hearing loss, or transient ischemic attack have been reported in some case reports (3-5). Headache is the most prevalent and generally the first symptom in patients with CVT, but various cohort studies represent that 5% to 30% of patients do not report headache at baseline (6). Evaluation of the characteristics of patients, who had CVT without headache, showed that these patients were older, and more often men. In these patients, paresis, seizures, isolated cortical vein thrombosis, brain parenchymal lesions, and malignancies were more frequent, and papilledema was less common (7).

We presented a case with acute analgesic and muscle-relaxant-resistant cervical pain and lateral bending. Thrombosis of right transverse and sigmoid sinus was detected in MRI and MRV of our patient. Downward extension of lateral sinus thrombosis into the sigmoid sinus and the proximal part of internal jugular vein could be the probable explanation of cervical pain in this case. Neck pain was reported as an infrequent consequence of the septic thrombosis of the external or internal jugular veins (8), and the internal vertebral venous plexus (9). Wasay et al. reported the presence of pain in the occipital and neck region in 17 out of 28 patients with involvement of sigmoid sinus alone or in combination with transverse sinus, due to inflammation and stretching of sinus walls (6). Besides, single or multiple cranial nerve palsies (III-VIII) without evidence of other signs and symptom have been reported in patients with thrombosis of the ipsilateral transverse/sigmoid sinus (10). To our knowledge, neck pain and lateral bending have never been reported as a sole symptom of transverse and sigmoid sinus thrombosis.

CT scan is often the first imaging for diagnosis of CVT in clinical practice; but it is normal in up to 30 percent of these patients, and most of the findings are not specific for CVT. However, in about one-third of patients, CT scan represents direct signs of CVT, which are the cord sign, and the empty delta sign. Brain MRI and MRV are the most sensitive imaging for detecting the thrombus in dural sinuses or veins (1).

Plasma D-dimer is a marker of endogenous fibrinolysis, and can be a good, available, and rapid diagnostic tool in suspected CVT patients (11), especially on emergency basis.

However, a negative D-dimer value does not rule out CVT.

The most frequent risk factors of CVT are prothrombotic conditions, either genetic or acquired, OCPs, pregnancy, puerperium, infection, malignancy, head injury, and mechanical precipitants (12). In more than 85 percent of adult patients with CVT, at least one risk factor can be detected. In the ISCVT cohort study on 624 adults with CVT, a prothrombotic condition was identified in 34 percent of all patients, and a genetic prothrombotic condition was detected in 22 percent of all patients (13). According to multiple observational studies, use of OCP increases the occurrence of CVT by 5 to 22 fold (14). The presentment of an inherited thrombophilia (such as Factor V Leiden, or prothrombin-gene mutation) enhances the odds further. Recent studies represented that the use of third generation OCPs are associated with an increased risk of CVT when compared to previous generation OCPs (15). We could not find any prothrombotic state, either genetic or acquired, in our patient, and OCP utilization and dehydration due to fasting was the only identifiable causes of her clot formation. Follow-up MRI, after three months of oral anticoagulation therapy, showed evidence of recanalization in our patient.

This case report suggested that in patients with intractable neck pain, especially those who are predisposed to conditions such as thrombotic complications by use of OCPs, CVT should be considered in the differential diagnosis, and appropriate neuroimaging and proper treatments must be carried out post haste. Finally, if this happens to women who take OCPs, it is recommended that they do not undergo estrogen-based therapies in the future.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

We thank Dr. Maryam Mashayekhi, the radiologist and MRI fellowship for assistance with quick report of neuroimaging, and providing patient's imaging.

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