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CASE REPORT

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Superior sagittal sinus thrombosis as unusual cause of headache: case report

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Abstract Headache is the most frequent symptom in patients with cerebral venous thrombosis. However, patients presenting with headache due to cerebral venous thrombosis are uncommon. The association between oral contraceptives and cerebral venous thrombosis is well known. We report the case of a young woman who was admitted to our department for sudden onset of headache. She had been taking oral contraceptives for 6 months. Early pharmacological approach with analgesics failed to alleviate symptoms. Magnetic resonance imaging (MRI) showed thrombosis of the posterior and middle thirds of the superior sagittal sinus (SSS). Because the patient was oligosymptomatic, medical treatment with high-dose heparin was started. A clinical follow-up showed headache regression after 2 weeks of therapy. Subsequent MRI showed partial recanalization of the SSS. The patient continued oral anticoagulants for 3 months. Eighteen months after discharge, the patient was symptom-free. We conclude that new, persistent or atypical headaches in patients taking oral contraceptives should be carefully evaluated for cerebral venous thrombosis.

Key words Cerebral venous thrombosis • Headache • Oral contraceptives • Heparin

Introduction

Cerebral venous thrombosis (CVT) is an unusual cause for severe headache [1]. Pathophysiologically it is characterized by a disturbance of the equilibrium between endogenous thrombogenic and fibrinolytic factors. In addition, the time course of CVT depends on the presence or absence of efficient venous collaterals. CVT has no single pattern of presentation, cannot be diagnosed on clinical grounds, and requires neuroimaging for diagnosis. Once diagnosis is established, a wide investigation should be carried out in search of the cause, and treatment started as soon as possible [2, 3].

In 1962 Lorentz [4] suggested an association between oral contraceptives use and the development of cerebrovascular diseases such as CVT. Since then, many cases have been reported in the literature and many studies have been performed on cerebral venous thrombosis in women taking oral contraceptives [5–7].

We report the case of a young woman taking oral contraceptives and suffering from severe headache due to superior sagittal sinus thrombosis successfully treated with heparin.

Case report

A 25-year-old woman was admitted to our department complaining of headache. Her headache had begun 15 days previously and it was described as a throbbing pain located in the

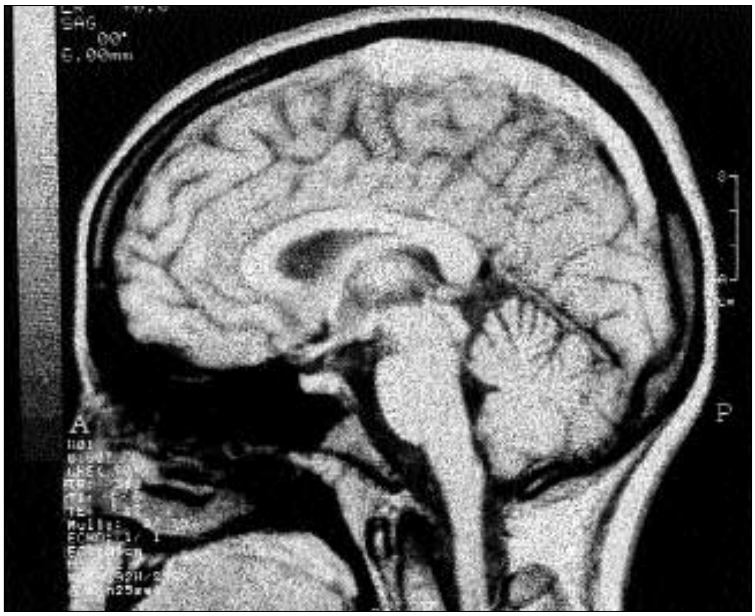

Fig. 1 Cerebral MRI with gadolinium enhancement. T1-weighted sagittal images showing a subacute thrombus in the posterior and middle thirds of the superior sagittal sinus

frontal and periorbital regions, of severe intensity, with no fluctuations during the day, exacerbated by the orthostatic position or by bending the head, sometimes with nausea and photophobia. The pain was refractory to oral analgesics. She denied fever, chills, sinus disease and visual problems. She had no previous history of migraine, nor did any family members. The patient had been taking oral contraceptives for a period of 6 months. There was no papilledema or sinus tenderness or other neurological symptoms or signs. Cerebral magnetic resonance imaging (MRI) with MR angiography revealed a thrombus, in subacute phase, in the middle and posterior portions of the superior sagittal sinus (Fig. 1). Routine workup for a hypercoagulable state (antithrombin III, proteins C and S, plasma homocysteine levels before and after oral methionine loading, anticardiolipin/antiphospholipid antibody, activated protein C resistance) revealed no abnormalities. Systemic medical treatment with high doses of heparin (30000 U/day with continous intravenous infusion) was started. Clinical follow-up showed headache regression after 2 weeks of therapy. One month later, MRI showed partial recanalization of the superior sagittal sinus. The patient was discharged symptom-free 22 days after admission. At home, she continued oral anticoagulants for 3 months. Eighteen months after discharge, the patient remained symptom-free.

Discussion

As reported by Bousser [8], CVT presents with different symptoms, and the mode of onset is also variable: it is subacute in 50% of cases, acute in 30% and chronic in 20%. A wide variety of clinical symptoms occur, and may be grouped into at least four main patterns (a) focal neurological deficits or partial seizures; (b) the pseudotumor cerebri syndrome; (c) subacute encephalopathy characterized mainly by a depressed level of consciousness and sometimes seizures without recognizable features of intracranial hypertension; and (d) a slowly progressive cavernous sinus thrombosis with a moderately painful third or sixth nerve palsy [8]. Nevertheless, many unusual presentations of headache cannot be classified into the previously mentioned patterns.

The presence of headache as the only symptom may be misleading, and may simulate other conditions such as subarachnoid hemorrhage, or even migraine. In different series, headache has been found in 70%–91% of patients with CVT, while the most frequent sign is papilledema, occurring in 27%–80% of patients [8–10]. The presence of headache, described by the patient as the first or the worst experienced, should lead the physician to suspect a secondary headache, such as headache in cerebral venous thrombosis.

The real incidence of intracerebral venous thrombosis is unknown because it goes unrecognized in many patients. It is likely to be lower than the incidence of approximately 1 per 1000 persons per year reported for deep-vein thrombosis. In 61% of affected women, intracerebral venous thrombosis occurs between the ages 20 and 35 years, which coincides with peak oral contraceptive use and incidence of pregnancy [2, 3, 11]. Using oral contraceptives is a well-known risk factor for CVT. In these women, there is a hypercoagulable state caused by increased platelet aggregation and adhesiviness, increased factors VII, IX and X and increased prothrombin and fibrinogen. The second important factor is changes in the vessel wall with endothelial proliferation [5–7].

Causes of, or predisposing conditions for CVT can be classified in infective and noninfective. The incidence of septic CVT has been reduced in developed countries since the use of antibiotics. Among the noninfective medical causes of CVT, congenital thrombophilia is the most frequent, particularly the increased resistance to activated protein C with factor V Leiden mutation and the 20210 G to A mutation of the prothrombin gene. Thrombosis secondary to severe local head injury is well known. Systemic lupus erythematosus, Behçet's disease and periarteritis nodosa are the rheumatologic diseases which are associated with CVT. Hypercoagulable states may be caused by nephrotic syndrome, antithrombin III deficiency, cancer and pregnancy. Cardiac diseases, such as congenital heart failure, cardiac insufficiency and pace maker use, may be also associated with CVT. CVT can be also caused by cirrhosis, Crohn's disease and ulcerative colitis [1, 8].

The diagnosis of CVT is possible with computed tomography or MRI. The former technique shows direct signs of CVT in one-third of cases. MRI allows the thrombus to be dated, since its signal features change with progression of the disease. In the hyperacute phase (0–24 hours), the thrombus is hypointense on T1-weighted images and hyperintense on T2-weighted images. In the acute phase (1–5 days), MRI shows hypointense images on both sequences. Later, the thrombus is hyperintense on both T1 and T2-weighted images in the subacute phase (5–14 days) and dishomogeneous after day 16. These changes depend on hemoglobin metabolism. Nevertheless, MRI does not correlate with disease prognosis, because signal anomalies can be also observed months after thrombosis, without correlation to clinical findings [12].

Treatment of CVT varies according to the clinical progression of the disease. Indeed, some physicians suggest conservative treatment and they reserve interventions only in patients with clinical deterioration. Stansfield, in 1942, first used heparin in a puerperal woman with focal neurological deficits secondary to venous thrombosis [13]. Since then, Einhaupl et al. [14] and De Brujin and Stam [15] performed the only two randomized studies on anticoagulant therapies in patients with cerebral venous thrombosis; they stated that heparin treatment is not only safe but also beneficial in these patients, even in cases with associated intracranial hemorrhage. Other physicians have advocated the use of fibrinolytic agents, with the goal of rapid clearance of the thrombus from the venous system. The development of selective catheterization allowed the successful treatment of many patients via transfemoral venous catheterism and direct instillation of thrombolytics (e.g. urokinase, streptokinase, rt-PA). However, the use of rt-PA seems to be correlated with the risk of minor hemorrhage or systemic coagulopathy risk. Selective fibrinolytic therapy is safe even in individuals with hemorrhage and nonhemorrhagic venous infarcts. Nevertheless, this treatment should be reserved to patients with fast clinical deterioration after the failure of systemic therapy with heparin [16].

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