

CEREBRAL THROMBOSIS OF THE TRANSVERSE SINUS

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ABSTRACT

Background and Aims: Cerebral venous thrombosis (CVT) is a rare disease associated with a extremely wide spectrum of clinical presentation, which often delays appropriate diagnosis. Transverse sinus thrombosis may be oligosymptomatic, presenting only with single or multiple cranial nerve lesions, which almost always initially lead to another diagnosis. The patients presenting with less severe and uncharacteristic symptoms were only exceptionally diagnosed until the advent of non-invasive brain imaging techniques.

Methods: We describe two patients with CVT, who presented with unusual clinical features, initially simulating a subarachnoid hemorrhage. They developed severe headache with acute onset, associated with vomiting, and neurological examination demonstrated papilledema and isolated peripheral sixth nerve palsy. We analyzed age, gender, personal history of infectious disorders, neuroradiological features, and response to medical treatment. CT brain scan was initially used and excluded cerebral hemorrhage or other brain lesion. MR venography identified thrombosis in the transverse sinuses in one patient, and in both superior sagittal and left transverse sinus in second patient. Intravenous heparin was infused.

Results: After a period of clinical improvement, the first patient presented a worsening of the bleeding and died from intracerebral frontal lobar hemorrhage. Blood flow was partially restored in the second patient; the MR venography showed partial recanalisation, by visualisation of slow flow in partially thrombosed sinuses, but he remained with severe visual impairment, secondary to papilledema.

Conclusion: CVT is an uncommon condition that often has severe clinical consequences. Early MR venography should be considered in patients with single or multiple cranial nerve lesions of uncertain etiology.

Key Words: transverse sinus thrombosis, cranial nerve lesions, MRI, complications.

INTRODUCTION

Cerebral venous thrombosis is an uncommon condition that often has severe consequences. Symptomatic thrombosis of intracranial sinuses and veins has been mainly observed in two groups of patients: subjects with bacterial or fungal infections, either systemic or localized to the cranium; subjects with noninfectious, miscellaneous conditions. In a substantial proportion (about 25%) of the patients with angiographically confirmed intracranial sinus venous thrombosis, there was no identifiable risk factor. The diagnosis of CST was infrequently made before 1970s.

One of the first descriptions of CVT is attributed to Ribes (1825), who reported the case of a patient who died six month after the onset of severe headache, epilepsy, delirium. Autopsy disclosed thrombosis of both the superior sagittal and transvers sinuses. Raymond (1880) gave the first description of cortical vein thrombosis. The clinical features of isolated cortical vein thrombosis are not well known. Loss of consciousness and intracranial hypertension may be exceptional in isolated cortical vein thrombosis.¹⁻⁴ CVT is an unfrequent condition that is extremely variable in its clinical presentation, mode of onset, imaging appearance, and outcome.

The classical description of CVT comprises headache, seizures, bilateral or alternating focal neurological deficits, in a variety of combinations and with different degrees of severity, with impairment of consciousness to progressive coma and death. The abnormalities described in autopsy of the brain with CVT are extremely variable.

Cerebral venous and sinuses thrombosis is a rare and severe disease that usually begins with severe headache, sometimes with sudden onset. Cerebral

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venous thrombosis may cause cerebral venous infarction, frequently hemorrhagic, that may lead to neurological deficits, epilepsy, or death. Obstruction of the sinuses often causes intracranial hypertension, which may produce papilledema of optic nerve and impaired vision. The etiologies are diverse, with over 100 causes identified; local causes such as trauma, tumors, infections, dehydration, Behcet disease, coagulopathies related to systemic disease, congenital coagulation disorders, pregnancy, post-partum period, use of oral contraceptives. The etiology remains unknown in 20% to 25% of the cases.^{1,5-10} The prognosis of CVT although much better than classically thought, remains largely unpredictable, with a wide discrepancy between clinical recovery and vessel recanalization. Many factors of poor outcome have been identified: extreme age, coma, severely raised intracranial pressure, infectious or malignant etiologies, hemorrhagic infarcts on CT, intercurrent complications such as seizures or pulmonary embolism. Ozawa¹¹ suggest as a rare complication after sinus thrombosis, the development of the dural-pial arteriovenous malformation.

The treatment of sinus thrombosis with heparin is controversial; heparin may arrest progression of thrombosis and prevent further infarction, but it may also cause hemorrhage in infarcted brain tissue. Low-molecular weight heparins, such as nadroparine, are as effective as unfractionated heparin, and cause fewer hemorrhagic complications. For now, heparine remains the first line treatment for CVT.^{4,7,12-14} Bruijn¹⁵ considers that anticoagulation is safe, even in patients with cerebral hemorrhage. The safety and efficacy of combined intrathrombus rtPA and intravenous heparin in CVT need further evaluation, because insufficient data are now available on efficacy and risks.^{16,17} The prognosis depends on the initial state and on treatment; the complete recovery ranges from 50%-80% in different case series, and the recurrence are reported in at most 10% of patients. In a number of a recent series mortality was ~ 10%.^{1,13}

The clinical symptoms of intracranial venous thrombosis vary with the location and the size of the occluded vessel. Clinical symptoms of CVT that involve the superior sagittal sinus, the transverse sinus and the large intracranial veins, differ from those observed in patients with cavernous sinus thrombosis. The mode of onset is also more variable and the spectrum of clinical presentation, extremely wide, as follows:

1. The most frequent syndrome is one of subacute or progressive onset of intracranial hypertension, and persisting, dull, generalized headache. Additional bilateral papilledema and small ventricles on CT, should

suggest a diagnosis of CVT; (a pseudotumor cerebri syndrome).

2. Another pattern of onset includes the sudden onset of focal neurological deficits; seizures and the neurological deficit that could not be related to a specific arterial territory, should suggest the same diagnosis;

3. A third pattern of clinical presentation is that of the subacute onset of multiple deficits, with or without signs of intracranial pressure.^{6,7,18} In some patients the extension of thromboses from the sagittal sinus to cortical veins leads to venous infarction. The infarction is most often bilateral, posterior parietal and hemorrhagic. Frank hematomas may develop. Brain edema and increased intracranial pressure can be fatal. Pulmonary embolism associated with sinus thrombosis, or from concurrent leg vein thrombosis, has been reported.^{1,2,13}

Bousser and Barnett (1992) suggested separating CVT into four groups: 1. with isolated intracranial hypertension; 2. with focal neurological signs; 3. with cavernous sinus syndrome; 4. with unusual presentation. The cranial nerve syndroms may be allocated to the last group. Ameri and Bousser reported the involvement of cranial nerves in 12% of all CVT cases, but cranial nerve palsies without other focal signs due to CVT were not observed.¹ Cranial nerve palsy in cerebral sinovenous thrombosis is rare, its pathophysiology remains unclear, and data from electrophysiological examinations in such patients are missing. Straub et al.¹⁹ report a case of extensive multiple CVT with isolated right peripheral facial palsy. Kuehen et al.² reported five cases of cranial nerves palsies, as the only features of isolated transverse sinus thrombosis.

PATIENTS AND METHODS

Two patients, both male, with intracranial hypertension syndrome and VI cranial nerve lesion, of undetermined origin, were studied. Presenting manifestations included seizures, severe headache, nausea and vomiting, papilledema and cranial nerve lesions. Both of them underwent full clinical examination, lumbar puncture, ECG, X-ray, CT and MRI. MRI was obtained with a 1-T whole body imaging system (Signa Horizon MR, General Electric, USA), using a circular polarized head coil. Each patient underwent a routine MRI protocol with T1-proton-density and T2-weighted axial images, including flash two-dimensional MR venography postprocessed by the maximum intensity projection algorithm and axial T1-weighted images after administration of gadolinium.

CASE HISTORIES

Case 1. A 55 year old man was admitted because of a 3 days history of repeated nausea, vomiting, headache and seizures. The patient was initially treated for suspected subarachnoid hemorrhage. Neurological examination revealed left VI cranial nerve palsy, intermittent diplopia and bilateral papilledema.

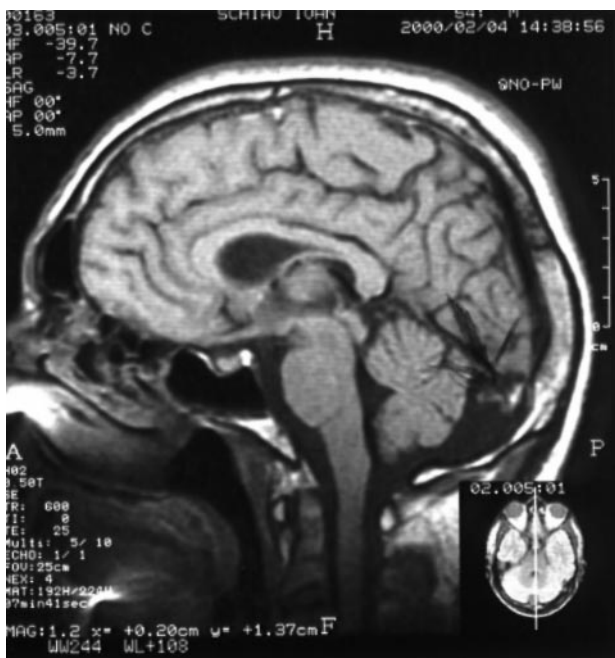
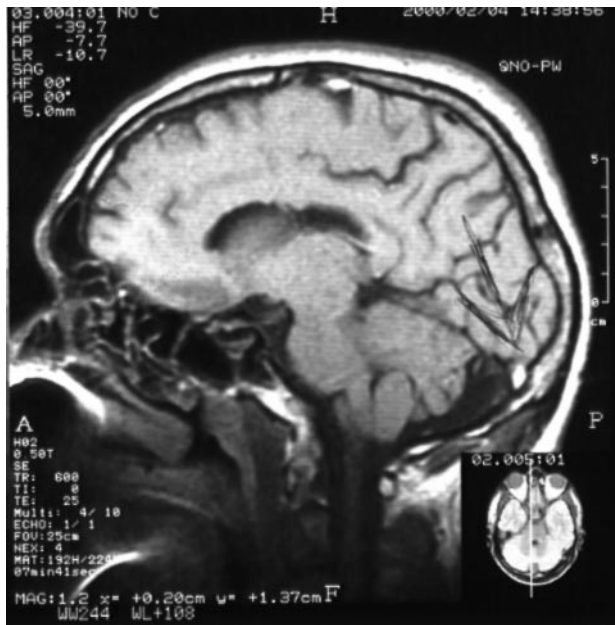


Figure 1. a,b. Case 1: sagittal cerebral MRI showing thrombosis in the territory of the superior longitudinal sinus, torcular, and to the junction with the left transverse sinus, a lesion of 2 cm with inhomogeneous density (arrow)

CSF was slight hemorrhagic. CT on the fourth day from onset, detected only a nonspecific hyperdensity in posterior fossa (cisterna magna). Only MR venography identified thrombi in superior sagittal sinus and at the junction of left transverse sinus and superior

sagittal sinus (Fig 1 and 2), associated with a cortical hemorrhagic component (Fig. 3), by spreading of thrombosis from sagittal sinuse to cerebral veins.

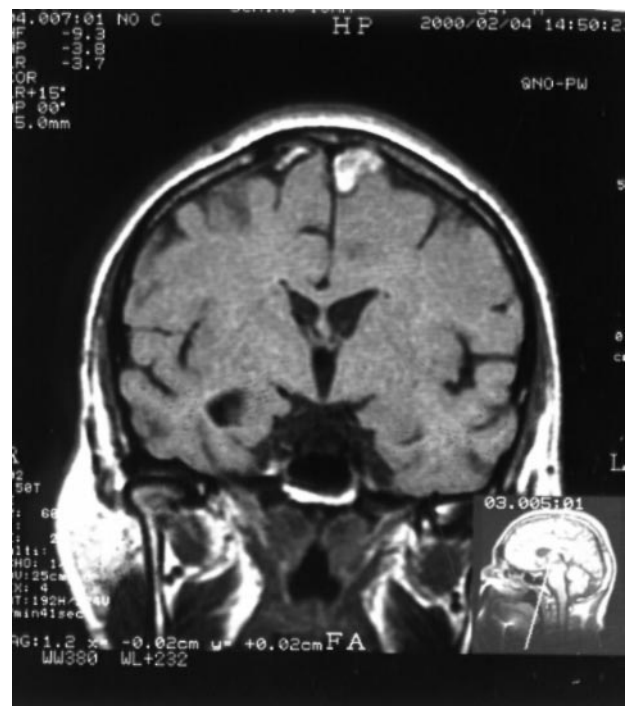


Figure 2. Case 1: coronal cerebral MRI showing extension of the thrombosis, mainly to the left side of longitudinal sinus

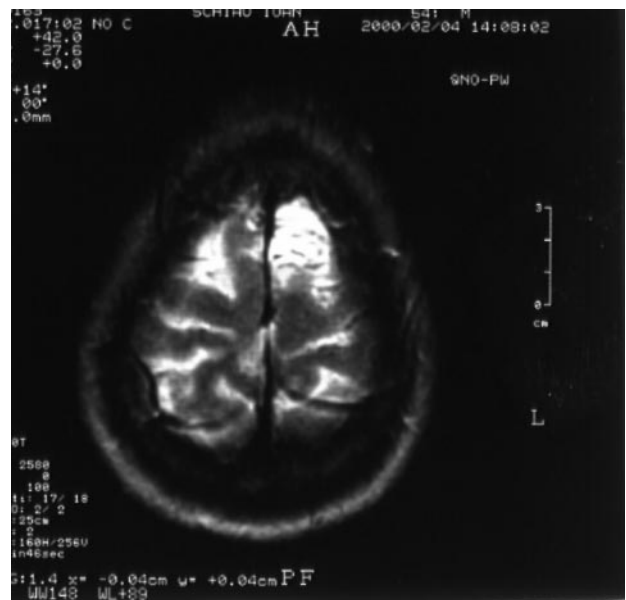


Figure 3. Case 1: cortical hemorrhagic component, from involvement of cortical veins

The patient received intravenous heparin for 4 days and nadroparine for the next 5 days; the symptoms improved, headache and left 6th cranial nerve palsy ceased, but after few days, suddenly hemorrhagic complication occurred. CT performed at that moment revealed hemorrhage (5/3 cm) in the frontal lobe and in the anterior interemispheric fissure, without focal deficit. On funduscopy, recent intraocular hemorrhages

were demonstrated. The patient died from intracerebral hemorrhage 25 days after admission.

Case 2. A 43 year-old- man was admitted after 2 weeks of repeated headache, which worsened despite analgesic treatment, nausea and vomiting, vertigo and imbalance, and discrete left ear pain for 2 days. At admission neurological examination demonstrated left periferal sixth cranial nerve palsy, nystagmus. CSF was normal, papilloedema was absent. Otoscopy showed no abnormality of ears or tympanic membrane. In his medical history there was an infection at his leg, five month before, and a facial Staphylococcal infection one month before. The episodes of vomiting, vertigo and imbalance stopped after three days, but headache worsened. Few days after the admission, intracranial hypertension developed, with slight bilateral papilloedema, progresive impairment of vision, and bilateral sixth nerves palsies. Brain CT scan was normal. Routine blood chemistry revealed no abnormalities. MR venography examination revealed a thrombosis of the bilateral transverse sinuses, more severe on the left, without associated brain lesions (Fig.4).



Figure 4. Case 2: cerebral MR venography showing bilateral thrombosis of transverse sinuses, with complete thrombosis of the left sinus

Intravenous heparin was immediately initiated: headache disappeared gradually, the sixth cranial nerves palsies resolved after 18 days, but the impaired vision remained, due to retinal damage secondary to papilloedema. MR venography performed after 16 days, showed partial recanalisation of bilateral transverse sinuses (Fig. 5).

RESULTS

Both patients were initially suspected of suffering

from other etiologies of cranial nerve lesions and papilledema. Neurological features and detailed history suggested the correct etiology, but MRI was needed to confirm it. The specific final diagnosis was established only after conventional MR venography was performed. Both of the patients were treated with heparin; the first patient received intravenous heparin for 4 days and nadroparine for the next 5 days. After several days of symptomatic improvement, he sustained hemorrhagic worsening. Flow restoration was incomplete in the second patient, with clinical improvement, except for visual impairment. The patient's headache and cranial nerve palsies resolved in conjunction with flow restoration, but he remained with persistent impaired vision. Optic nerve decompression was not performed to relieve intracranial pressure.



Figure 5. Case 3: control MR venography after 16 days, showing partial recanalisation, mainly in the left transverse sinus, with segmental occlusion of right transverse sinus and persistence of a residual thrombus in the torcular

DISCUSSIONS

Cerebral venous and sinuses' thrombosis is a rare and severe disease that usually begins with severe headache. Sinus thrombosis may cause cerebral venous infarction, which are frequently hemorrhagic and may lead to neurological deficits, epilepsy, or death. Obstruction of the sinuses often causes intracranial hypertension, which may result in optic nerve papilloedema and impaired vision.

Patients presenting with less severe and uncharacteristic symptoms, were only exceptionally diagnosed until the advent of non-invasive brain imaging techniques.

Computerized tomography was initially used to identify sagittal sinus thrombosis. More recently MRI

has improved the diagnostic sensitivity by simultaneous visualization of the affected brain tissue and the venous system, also demonstrating the capability of identifying transverse sinus thrombosis in the posterior fossa. The present “gold standard” for the diagnosis of CVT is MRI and MR venography.^{1,2,13,19-21} More recent reports describe patients presenting with a history of cranial nerve lesions as the major neurological findings accompanied by mild cerebellar incoordination and papilledema, suggesting cerebellar infarction or posterior fossa tumour.

Our first case presented with headache, papilledema, 6th nerve palsy, and MRI showed transverse sinus thrombosis in combination with obstruction of superior sagittal sinus, and cortical hemorrhage. Intracerebral hemorrhage is a well-known complication of CVT, being also the cause of his death. Depending on the individual anastomoses of the ventral venous network, thrombosis of the lateral sinuses can produce a venous overload, “venous congestion”. Venous congestion is different from venous infarction, and is associated with reduced venous drainage and vascular oedema. Oedema is probably the most common abnormality responsible for neurological signs, in addition to other parenchymal abnormalities including hemorrhage and ischemia. The hemorrhagic worsening in the first patient was probably accelerated by anticoagulation and this may be the mechanism of death for our first patient.

The second case, presented with headache, vertigo, nystagmus, 6th nerve palsy, and after few days, intracranial hypertension, papilledema, and finally, amaurosis; MRI showed in this case, bilateral transverse sinus thrombosis, more pronounced on the left. He progressively developed a syndrome of subacute intracranial hypertension, and visual impairment was secondary to papilledema.

History and clinical features were initially uncharacteristic for the transverse sinuses lesions. Straub et al¹⁹ and Kuehnen et al² reported multiple cranial nerve palsies (VII, VIII, III, V, VI) in transverse sinus thrombosis. In our cases, sixth nerve palsy proved that increased intracranial pressure was present.

CONCLUSIONS

- Cerebral venous thrombosis may affect one or more cerebral sinuses, cortical veins, or both structures.
- Early diagnosis of cerebral transverse sinus venous thrombosis is often difficult because of

nonspecific and heterogeneous clinical presentation.

- The prominent symptom of our patients was intracranial hypertension, associated with 6th nerve palsy.
- For a definitive diagnosis MRI and MR venography are indispensable.
- Prognosis remains largely unpredictable.
- Although the data for heparin therapy in CVT are generally favorable, unfortunately, definitive conclusions about safety and efficacy of intravenous heparin in patients with CVT cannot be drawn.

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