Isolated Lateral Sinus Thrombosis
A Series of 62 Patients

Mariem Damak, MD; Isabelle Crassard, MD; Valérie Wolff, MD; Marie-Germaine Bousser, MD

Background and Purpose—Isolated lateral sinus thrombosis was long considered a complication of middle ear disease. Little attention has been recently paid to this variety of thrombosis. We therefore reviewed all cases of isolated lateral sinus thrombosis prospectively collected in our center (1997 to 2006).

Methods—Among 195 patients with cerebral venous thrombosis (CVT), we identified 157 patients with lateral sinus thrombosis, including 62 patients with isolated lateral sinus thrombosis. Clinical, etiologic, and prognostic features were compared with those of other 133 CVT cases.

Result—Sixty-two patients (32%) had isolated lateral sinus thrombosis. Headaches were present in 95% of patients. The main clinical presentation was isolated headache in 28 patients (45%), whereas 15 (24%) had isolated intracranial hypertension. Nineteen patients (31%) had at least one focal sign (deficit and/or focal seizure). Dysphasia was the most common one (8 patients). Compared with the other 133 CVT cases, presentation with isolated headaches was the most frequent one ($P<0.001$). Parenchymal lesions were found in 19 cases and were less frequent than in other CVT cases ($P=0.007$). Numerous causes or predisposing factors were identified without a difference in repartition with other patients with CVT, particularly for local or infectious causes. Treatment consisted of anticoagulation in all patients. Clinical outcome was good with complete recovery in 57 patients (92%). One patient died after the occurrence of massive pulmonary emboli despite adequate anticoagulation. Three patients had sequelae (no difference with the other CVT).

Conclusion—Isolated lateral sinus thrombosis is a frequent variety of CVT, accounting for one third of all CVT. It presents in more than two thirds of cases with headache as the only symptom either isolated or less frequently associated with papilledema. This stresses the need for CVT workup in a patient with a recent unexplained headache. (Stroke. 2009;40:476-481.)

Key Words: headache lateral sinus sinus thrombosis

Cerebral venous thrombosis (CVT) is an infrequent condition characterized by a wide spectrum of clinical presentations and modes of onset, which depend on the site, extent, and rate of progression of thrombosis.$^{1,2}$ The 2 most frequent sites of thrombosis are the superior sagittal sinus (SSS), affected in 62% to 80% of cases, and the lateral sinus (LS) involved in 38% to 86% of cases.$^{1,3-5}$ A remarkable feature of CVT is that, in approximately 75% of cases, thrombosis affects several sinuses, the most frequent combination being SSS + LS.$^{1}$ The isolated involvement of one sinus is thus rare, less than 30% for SSS and 10% for the LS.$^{1,3-4}$

Isolated LS thrombosis (LST) has mostly been reported in otologic series$^{6-8}$ as a consequence of acute or chronic ear and mastoid infections, hence the term “otic hydrocephalus” coined by Symonds when LST manifested as raised intracranial pressure.$^{9}$

Because of, first, the dramatic reduction of intracranial complications of infectious ear diseases since the antibiotic era, and second, the possibility of early diagnosis of CVT since the widespread use of neuroimaging, the pattern of presentation of isolated LST is likely to have changed. We describe here the main clinical and MRI features, the causes and outcome of all patients with isolated LST prospectively collected in our center in the past 10 years, and compare them with those of the other varieties of CVT examined during the same period.

Patients and Methods

The present study is based on a prospective cohort of 195 consecutive patients with CVT admitted to our department between September 1997 and April 2006.

Diagnosis of CVT was based on MRI combined with MR venography and/or helical cerebral CT venography. The diagnosis of LST was based on the association of negative (nonvisualization of the entire LS or of its sigmoid part at CT angiography or MR venography) and positive signs (definite spontaneous LS hyperdensity on nonenhanced CT or hypersignal on MRI T1 or T2 WI or hyposignal on MRI T2*SW)$^{10,11}$ (Figure 1). The presence of parenchymal lesions was assessed on both nonenhanced CT scan and MRI (T1, T2, fluid-attenuated inversion recovery, diffusion, and T2*SW).
LST was defined as isolated when, at admission in our department, there was no associated SSS, deep venous system, or straight sinus thrombosis. Patients with cortical vein thrombosis were also excluded except when the vein afferent to the thrombosed LS was the only cortical vein involved.

After January 2001, mastoid air sinus abnormalities consisting of increased T2 WI MRI signal in the mastoid air space were systematically looked for and recorded.

Symptoms and signs, presence or absence of papilledema, modes of onset, neuroimaging findings, causes and risk factors, including a thrombophilic workup, and outcome at discharge and at 1 year were systematically recorded. The onset was defined as acute (≤2 days), subacute (2 days to 1 month), or chronic (over 1 month). All patients were treated acutely by a therapeutic dose of intravenous heparin or low-molecular-weight heparin. Disability at discharge and at 1 year was classified according to the modified Rankin Scale (mRS) as complete recovery (mRS 0 to 1), partial recovery (mRS 2), dependent (mRS 3 to 5), or death (mRS 6).

Statistical Analysis
Patients with isolated LST were compared with the rest of CVT cohort using the χ² test for dichotomous data and Student t test for continuous data. All significance levels reported were 2-sided and a probability value of 0.05 was considered to indicate statistical significance.

Results
Baseline Clinical Characteristics
Among the 195 patients with CVT, 108 (55%) had SSS thrombosis and 157 (80%) had LST. The present study is based on 62 (32%) who had isolated LST and who were compared with the other 133 patients.

Thrombosis affected the left LS in 36 patients (58%), the right LS in 25 (40%), and both in one patient (2%).

The female: male sex ratio (4:1) and the mean age (38 years) were the same in the 2 groups. The onset was more frequently subacute (80%) or chronic (10%) and less frequently acute (10%) than in other CVT.

Presenting symptoms and signs are listed in Table 1. Headache was in both groups by far the most frequent presenting symptom (95% and 96%, respectively), but papilledema was less frequent (22%) in isolated LST than in other CVT (47%). Seizures, dysphasia, and cranial nerve palsies were far less frequent in isolated LST than in other CVT. By contrast, the frequency of cerebellar ataxia was the same (13%) in both groups. Other signs such as cerebellar ataxia, hearing loss, vertigo, and VIth or VIIth cranial nerve palsy were rare in both groups.

The main patterns of presentation are listed in Table 2. Nearly half the patients (45%) with isolated LST presented with headache only, far more than in patients with other CVT (9%). A focal syndrome with focal deficits and/or seizures was less frequent in patients with isolated LST (31%) than in patients with other CVT (63%). Roughly one fourth of the patients in both groups had isolated intracranial hypertension.

Neuroimaging
Initial noncontrast CT scan was performed on an emergency basis in 57 patients (92%). It was normal in 23 cases (40%).
It showed definite spontaneous hyperdensity of the lateral sinus in 26 patients (45%; Figure 1). Other abnormalities were identified in 15 patients: parenchymal hypodensity in 6, parenchymal hyperdensity or mixture of hypo- and hyperdensity in 8, associated with sulcal hemorrhage in 2, isolated sulcal hemorrhage in one.

MRI with MR venography was performed in 58 patients (94%) showing by definition the positive and negative signs of LST. Other abnormalities were parenchymal lesions in 18 (31%) mostly in the temporal lobe (80%; Figure 2). Ten were nonhemorrhagic and 8 were hemorrhagic, including 2 associated with sulcal hemorrhage. One patient had isolated sulcal hemorrhage. Overall, patients with isolated LST had less neuroimaging lesions (31%) than patients with other sites of CVT (51%; \( P = 0.007 \)). Mastoid air sinus abnormalities on MRI T2 WI, prospectively looked for in 39 patients, were present in 11 patients (28%) invariably ipsilateral to LST (Figure 2C).

**Clinicoradiological Correlations**

Among the 28 patients with isolated headache as the initial presentation, 17 had a left LST and 11 a right LST. All but 2 (93%) had otherwise normal neuroimaging examinations. One had an isolated sulcal hemorrhage visible on both CT scan and MRI fluid-attenuated inversion recovery imaging. The other had a small parenchymal lesion on MRI fluid-attenuated inversion recovery imaging in the left temporal lobe.

In the 15 patients with isolated intracranial hypertension, only one had bilateral LST. In the 14 others, LST was unilateral, on the left side in 6, on the right in 8, and it was associated with contralateral hypoplasia and/or extension to the torcular. Three patients had small parenchymal involvement consisting of right temporal lesions in 2 and left temporal in one.

Among the 19 patients with focal deficits or seizures, 13 had a left LST and 6 a right LST. A parenchymal lesion was detected in 14 of them (74%), mostly in the temporal lobe.

**Causes and Predisposing Factors**

Causes and predisposing factors are summarized in Table 3. No significant difference between isolated LST and other CVT was found regarding causes and risk factors, even when considering local causes (6% versus 10%). Only one patient with isolated LST had otitis media, 2 had tonsillar abscesses, one had a homolateral cholesteroloma, one had a homolateral bone hemangioma, and one had LST after surgery for cervical syringomyelia.

**Treatment and Outcome**

Besides anticoagulation, additional treatments were prescribed according to the clinical presentation and causes: antiepileptic drugs in patients with seizures and acetazolamide and/or lumbar puncture in patients with isolated intracranial hypertension. The 3 patients with local infection received antibiotics; one patient had surgical treatment for sinusitis.

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**Table 2. Patterns of Presentation in Patients With Isolated LST versus Other Sites of CVT**

<table>
<thead>
<tr>
<th></th>
<th>Isolated LST (n=62)</th>
<th>Other CVT (n=133)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Isolated headache</td>
<td>28 (45)</td>
<td>12 (9)</td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>Isolated intracranial hypertension</td>
<td>15 (24)</td>
<td>36 (28)</td>
<td></td>
</tr>
<tr>
<td>Focal syndrome=encephalopathy</td>
<td>19 (31)</td>
<td>83 (63)</td>
<td></td>
</tr>
<tr>
<td>Diffuse encephalopathy</td>
<td>0 (0)</td>
<td>19 (14)</td>
<td></td>
</tr>
</tbody>
</table>

**Table 3. Causes and Risk Factors in Patients With Isolated LST versus Other Sites of CVT**

<table>
<thead>
<tr>
<th></th>
<th>Isolated LST (n=62) (%)</th>
<th>Other CVT (n=133) (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Local causes (infectious or not)</td>
<td>6 (10)</td>
<td>8 (6)</td>
</tr>
<tr>
<td>Gyneco-obstetric causes</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Current oral contraceptive use</td>
<td>27 (54)</td>
<td>52 (49)</td>
</tr>
<tr>
<td>Pregnancy or puerperium</td>
<td>3 (6)</td>
<td>18 (17)</td>
</tr>
<tr>
<td>General diseases, coagulation disorders</td>
<td>40 (64)</td>
<td>70 (52)</td>
</tr>
<tr>
<td>Coagulation disorders, thrombophilia</td>
<td>25</td>
<td>35</td>
</tr>
<tr>
<td>Systemic disorders</td>
<td>17</td>
<td>26</td>
</tr>
<tr>
<td>Cancer and malignant hemopathies</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>More than one cause or risk factor</td>
<td>23 (37)</td>
<td>34 (26)</td>
</tr>
<tr>
<td>None identified</td>
<td>8 (13)</td>
<td>17 (13)</td>
</tr>
</tbody>
</table>

*Patients may have more than one risk factor.
Table 4. Clinical Outcome at Discharge and at 1-Year Follow-Up

<table>
<thead>
<tr>
<th></th>
<th>Isolated LST (n=62)</th>
<th>Other CVT (n=133)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>At Discharge</td>
<td>At 1 Year</td>
</tr>
<tr>
<td>Complete recovery (mRS 0–1)</td>
<td>53 (85%)</td>
<td>57 (93%)</td>
</tr>
<tr>
<td>Partial recovery (mRS 2)</td>
<td>5 (8%)</td>
<td>2 (3%)</td>
</tr>
<tr>
<td>Dependent (mRS 3–5)</td>
<td>3 (5%)</td>
<td>1* (2%)</td>
</tr>
<tr>
<td>Death (mRS 6)</td>
<td>1 (2%)</td>
<td>1 (2%)</td>
</tr>
</tbody>
</table>

*mRS=3.

tonsillar abscess. One patient underwent a temporal hemato-
toma evacuation on an emergency basis because of a severe mass effect.

Disability at discharge and at 1 year is indicated in Table 4. The overall prognosis was good with 85% of patients having an mRS of 0 or 1 at discharge. This was significantly better than for the other varieties of CVT. At 1 year, there was no difference between the 2 groups with, respectively, 92% and 91% of patients with an mRS of 0 to 1.

Among the 62 patients with isolated LST, 5 had at discharge an mRS of 2 and 3 and had an mRS of 3 to 5. At 1 year, 2 patients had an mRS of 2 because of diminished vision and one an mRS of 3 because of motor deficit and dysphasia. One patient died during the acute phase. She worsened rapidly after admission and developed temporal hematoma. Local thrombectomy was performed but the patient did not regain consciousness. A decompressive craniectomy was then performed. The patient started to improve but 6 days later, she died of massive pulmonary emboli despite adequate anticoagulation.

Discussion

In this prospective series of 195 patients with CVT seen in the last 10 years in our institution, LS was the most frequently involved sinus (80% versus 55% for SSS) and it was also the most frequently involved in isolation, ie, without other sinuses involved (32% versus 9% for SSS). This high frequency of LST compared with SSS thrombosis$^{4,13,14}$ raises the question of LST diagnosis. We think overdiagnosis of LST in our study is unlikely because LST diagnosis was based on the association of the visualization of the thrombus itself (hyperdensity on CT scan, hypersignal on MRI T1 and T2, hyposignal on MRI T2*) and the nonvisualization of the entire LS or its sigmoid part on angiography (MR venography, CT angiography, conventional angiography).$^2$ The 2 main diagnostic pitfalls of LS, ie, LS hypoplasia and arachnoid granulation, have thus been carefully ruled out. LS hypoplasia can indeed be mistaken for LST if the diagnosis is based purely on the absence of flow on angiography but not if the positive image of the thrombus is detected in the nonvisualized vessel.$^{11}$ Similarly, arachnoid granulations, particularly when they are large, can be misinterpreted as a focal thrombosis if the diagnosis is based purely on the presence of a “positive” image within the sinus but not if there is an absence of flow in the entire LS or its sigmoid part.$^{14}$

Our series of isolated LST differs from previous large series, which were collected in ear, nose, and throat departments$^{6–8,15–18}$ where LST most frequently occurred as a complication of otitis media or other local infection.$^7,8,15–21$ The typical pattern was that of patients with an old or recent history of discharging ears who had neglected otitis media and presented with retroauricular swelling, otorhrea, fever, headache, nausea, and vomiting. In old series, patients were treated by internal jugular vein ligation$^{19,20}$ and, in more recent ones, by antibiotics and eventually surgery.$^7,21$ In our series, only 3 patients had a local ear, nose, and throat infectious cause (one otitis media, 2 tonsillar abscesses), not more than other varieties of CVT. This low rate of local infectious causes is consistent with recent series coming from countries where antibiotics are widely used.$^3,13$ A recruitment bias is unlikely because our institution has the only ear, nose, and throat emergency department for adults open 24 hours for the whole Paris area. Although today rare, local causes—infectious or not—should be systematically looked for by an ear, nose, and throat examination in patients with LST.

When comparing isolated LST with other CVT, there was no difference regarding age, sex, and the main causes; the main difference pertains to the clinical presentation with more isolated headache and less encephalic signs. Headache was the only sign in 45% of patients with isolated LST compared with 10% in other CVT. This unusually high rate of isolated headache might be partly due to the presence in our institution of an emergency headache center recruiting 8000 patients per year and allowing prompt investigations for patients with unexplained recent headache.$^{22}$

Among patients with isolated headache, 94% had a normal brain MRI. The exact mechanism of headache in such patients who have no intracranial hypertension, subarachnoid hemorrhage, menigitis, or intracranial lesion remains unknown. A local inflammatory reaction is a possibility with dilatation of vessels in the sinus walls as suggested by the frequent contrast enhancement surrounding the clot, known for SSS thrombosis as the “empty delta sign,” but also present in LST.$^{22}$ The pain might also be due to the irritation or stretching of nerve fibers in the walls of the occluded sinus, which often becomes round with a bulging aspect.$^{23}$

Isolated intracranial hypertension resumed the clinical presentation in approximately one fourth of patients with isolated LST as well as with other CVT. Such presentation is one of the most classical ones in CVT, particularly when several sinuses are occluded without involvement of cerebral veins.$^{13,24,25}$ In our series, it was mostly observed when the thrombosed LS was the dominant one, with hypoplasia of the other, or when thrombosis involved the torcular or, in one case, both LS.

Encephalic signs such as seizures, focal deficits, and disorders of consciousness were less frequent (one third) in isolated LST than in other CVT (two thirds). These differences in presentation are related to a significant difference in the frequency of brain lesions: 31% in isolated LST versus 51% in other CVT. The main focal sign was dysphasia due to a left temporal ischemic or hemorrhagic lesion secondary to...
thrombosis of the vein of Labbé, which drains in LS. Its frequency (13%) was the same as in other CVT. Motor deficit (3%) was 10 times less frequent than in other CVT and seizures (16%) 3 times less. No patient presented with diffuse subacute encephalopathy. Previous reports suggest that LST can manifest as isolated or multiple cranial nerve palsy.\textsuperscript{26,27} This was rare in our series; one patient had facial palsy and 2 had hearing loss, adding to a previous case report.\textsuperscript{28} Hearing loss may be due to the extension of thrombosis to cochlear veins, which empty into the lateral sinus directly or through the inferior petrosal vein.

Regarding imaging, attention has recently been drawn to the possibility in patients with LST of ipsilateral mastoid abnormalities with increased T2-weighted signal in the mastoid air spaces, often with a trabecular pattern.\textsuperscript{12} Such abnormalities were present in 39% of patients with LST in Fink’s original description and in 28% in our series, but not in the 3 patients who had an infectious local cause. When present, they should not be mistaken for mastoiditis. They may also help to differentiate LST from LS hypoplasia. They are due to mucosal edema and effusion, likely secondary to venous congestion as a consequence of LST. They all resolved during follow-up (MRI at 3 months).

The outcome of patients with isolated LST was better at discharge than that of patients with other CVT, but, at 1 year, there was no significant difference between the 2 groups.

Although the overall prognosis of isolated LST is good, this condition cannot be considered benign because one patient was still dependent at 1 year because of motor deficit and dysphasia and another patient died during the acute phase. Although at entry this patient who just presented with headache and dysphasia had none of the recognized factors of poor outcome,\textsuperscript{2} she rapidly became stuporous because of worsening of intracranial hemorrhage and development of massive brain edema. Local thrombectomy failed, decompressive craniectomy was performed, but she died of pulmonary embolism. This case illustrates that, on an individual basis, prognosis remains largely unpredictable and that, despite anticoagulation, pulmonary embolism remains a major cause of death in patients with CVT.

It also shows that, although the majority of patients with isolated LST as well as with other CVT do well with conventional treatment based on heparin, a few patients require aggressive treatment such as decompressive surgery to prevent death from herniation.\textsuperscript{29,30}

In summary, thrombosis limited to LS (with or without involvement of tributary veins) accounts for one third of all CVT in our series. Bilateral LST, without involvement of other sinuses is rare, only one of 62.

Isolated LST does not differ from other CVT regarding age, sex, and causes. In particular, it is today, at least in adults, rarely secondary to an ear, nose, and throat infection. Like in other CVT, the overall prognosis is good with 92% of complete or nearly complete recovery at 1 year, but management may eventually require decompressive surgery. Pulmonary embolism remains a cause of death, even in patients treated with heparin.

The most important finding in this series is that more than two thirds of patients had headache as the only symptom during the whole course of the disease. Only a minority of such patients had papilledema so that altogether, isolated headache was the clinical presentation in 45% of cases. This stresses the need to systematically look for LST (and other CVT) in patients with recent headache even in the absence of associated symptoms and signs. Positive imaging of the thrombosis itself together with the nonvisualization of the sinus involved at MRI/MR angiography or CT/CT angiography are required to diagnose LST and rule out LS hypoplasia and arachnoid granulations, which could be mistaken for LST. Although some patients may recover spontaneously,\textsuperscript{1} heparin remains the treatment of choice in isolated LST as well as in other varieties of CVT.\textsuperscript{2,31}

Disclosures

None.

References


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