

## Ipsilateral Dural Thickening and Enhancement: A Sign of Isolated Cortical Vein Thrombosis? A Case Report and Review of the Literature

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### Key words

- Ipsilateral dural thickening
- Isolated cortical vein thrombosis
- Magnetic resonance imaging

### Abbreviations and Acronyms

**CSF:** Cerebrospinal fluid  
**CT:** Computed tomography  
**DSA:** Digital subtraction angiography  
**EA:** Epidural anesthesia  
**GE:** Gradient echo  
**ICVT:** Isolated cortical vein thrombosis  
**LP:** Lumbar puncture  
**MRI:** Magnetic resonance imaging  
**SVT:** Sinus vein thromboses  
**SWI:** Susceptibility-weighted imaging  
**UFH:** Unfractionated heparin

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### INTRODUCTION

Isolated cortical vein thrombosis (ICVT) is a rare condition found in approximately 6% of all intracranial veins thromboses, mostly in conjunction with sinus vein thromboses (SVTs).<sup>1</sup> The highest risk factor, with an occurrence rate of 35%, for developing ICVT is pregnancy and puerperium. Only 116 cases of ICVT have been reported in the literature.<sup>2</sup> Typical symptoms are headaches (71%), seizures (58%), and focal neurologic deficits (62%). The mean time between the appearance of symptoms and diagnosis is 7 days.<sup>2</sup>

Diagnosing ICVT on noninvasive cerebral images is challenging because of the high variability of cortical veins and the difficulty in identifying small occluded vessels. Detection of ICVT on native computed tomography (CT) scans refers to a linear hyperdensity that has been described as the

■ **BACKGROUND:** Isolated cortical vein thrombosis (ICVT) being a rare condition (6% of intracranial vein thromboses), no clinical guidelines and few radiologic clues to it have been established. ICVT mostly appears in conjunction with sinus vein thromboses. ICVTs primarily occur during pregnancy and puerperium (35%). The great variability of cortical veins and difficulty identifying small occluded vessels complicate ICVT diagnosis. We present the first case of isolated ipsilateral dural thickening and enhancement as a potential radiologic sign of ICVT shown on magnetic resonance imaging.

■ **CASE DESCRIPTION:** A 30-year-old woman presented with sudden position-independent severe headache and neck pain 2 weeks postpartum. Standard magnetic resonance imaging revealed ipsilateral dural thickening and enhancement of the meninges (left hemisphere). The symptoms and findings were interpreted as a post-lumbar puncture syndrome associated with the epidural anesthesia during labor and birth. Deteriorating, the patient was referred to our hospital after a computed tomography scan had revealed atypical left parietal intracranial hemorrhage. Digital cerebral subtraction angiography confirmed a left parietal ICVT as the underlying disease. When systemic anticoagulation was initiated, the patient's condition further deteriorated. Progressive aphasia and right-sided face and arm weakness and numbness developed as a result of increased intracranial hemorrhage volume. We therefore performed craniotomy and hematoma evacuation, after which the patient fully recovered.

■ **CONCLUSIONS:** We hypothesize that ipsilateral dural thickening and enhancement in patients presenting with severe headache may indicate ICVT and we suggest further diagnostic workup, using venographic study such as computed tomographic venography or magnetic resonance venography. In an inconclusive venographic study with high clinical suspicion for ICVT, catheter cerebral angiography is indicated.

cord sign, which is an indication of changes in blood density within the thrombosed vein.<sup>3,4</sup> Typical signs of ICVT on magnetic resonance imaging (MRI) are signal loss of the affected cortical vein in T2-susceptibility-weighted gradient echo (GE) sequences, similar in shape to the cord sign of cortical vein thrombosis on CT scan. Various abnormalities in parenchymal signal intensity may also be found close to the occluded vein.<sup>1</sup> Digital subtraction angiography (DSA) is still considered the gold standard to confirm an ICVT diagnosis when radiologic (CT scan or MRI) or clinical examinations produce suspicious

results.<sup>1</sup> However, there is increasing evidence that MRI with gadolinium-enhanced T1 three-dimensional GE sequences and heavily susceptibility-weighted imaging (SWI) are the most sensitive diagnostic tools for the detection of ICVT.<sup>5,6</sup>

It is well documented that dural puncture (eg, lumbar puncture [LP] or epidural anesthesia [EA] with cases of accidental dural puncture) can lead to intracranial hypotension through the leakage of cerebrospinal fluid (CSF), with typical changes on MRI (diffuse image) and bilateral pachymeningeal enhancement (up to 83% of patients with CSF hypovolemia).<sup>7</sup> No cases of

unilateral dural enhancement as a sign of intracranial hypotension have been reported. However, incidental occurrence of ICVT on CT and MRI scans after LP, including a cord sign of the affected vein and intraparenchymal lesion, have also been described.<sup>8</sup> One of the hypothesized pathophysiologic causes of cortical vein thrombosis is intracranial hypotension, which causes a downward movement and kinking of the veins and might lead to thrombosis.<sup>1,3,8</sup> To the best of our knowledge, we present the first case of ipsilateral dural thickening and enhancement caused by ICVT. Differential diagnosis, potential pathogenesis, and the diagnostic workup of ICVT are discussed.

### CASE REPORT

A 30-year-old woman who had delivered a healthy baby under EA presented 2 weeks postpartum with sudden position-independent severe headache and neck pain without any focal neurologic deficit. Standard MRI with fluid-attenuated inversion recovery without SWI and gadolinium-enhanced T<sub>1</sub> three-dimensional GE sequences revealed no sign of intracranial bleeding, sinus venous thrombosis, or other pathologic findings, apart from left hemispheric dural thickening and enhancement (Figure 1B, C). This had not been evident on MRI performed during examinations into the cause of her migraines a year earlier (Figure 1A). The symptoms and the new radiologic findings were interpreted as post-LP syndrome induced by CSF

hypovolemia. This had possibly been caused by an accidental dural puncture during EA administered during labor and birth. The patient was sent home and prescribed analgesic drugs and bed rest. The headache persisted despite the medication and the patient had a seizure.

The patient was found unconscious at home 4 days after the MRI. During the transfer to the hospital, she suffered a tonic clonic seizure lasting for 30 seconds. The patient presented at the referring hospital with a Glasgow Coma Scale score of 6 and isocoric pupils with a slower reaction to light on the left side. On admission, a CT scan showed a 1.7 × 3.4 cm left parietal intracranial hemorrhage (ICH). Levetiracetam antiepileptic treatment was administered and the patient was referred to our hospital for further diagnosis and treatment.

The CT scan showed left parietal ICH as well as a cortical vein cord sign (Figure 2). After reviewing the CT scans and previous MRI results, ICVT was suspected but the intracerebral bleeding raised doubts that the CT hyperdensity was sufficient to confirm the diagnosis. Cerebral DSA was performed to elucidate the cause of bleeding. It showed no cerebral arteriovenous malformation or dural fistula and no direct sign of SVT. Instead, we found an irregularity in the contrast phases in the left parietal lobe and a missing cortical vein as well as a delayed irregular drain, compared with the opposite side (Figure 3). As a result, delayed ICH caused by ICVT was

diagnosed. Focal dural thickening and dural enhancement in MRI were located exactly in the region of the suspected ICVT.

The patient was subsequently monitored in our stroke unit and therapeutic anticoagulation with unfractionated heparin (UFH) was initiated, but her condition deteriorated. She developed incomplete aphasia, incomplete right-sided face and arm weakness and numbness, and an increasingly severe headache. A follow-up CT scan revealed a slight increase in bleeding, and conservative treatment was continued. During the course of the day, the patient became somnolent and developed complete right-sided face and arm weakness and numbness as well as global aphasia. A further CT scan showed a progression of the ICH (4.1 cm × 6 cm). Because of the size of the ICH, a craniotomy and microsurgical clot removal were indicated at this point.

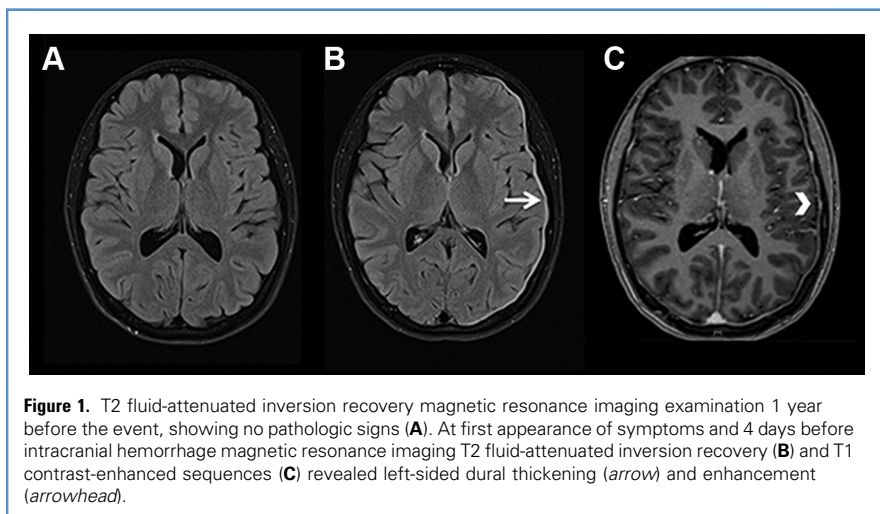
There were no intraoperative complications. The thrombotic cortical vein was visualized clearly and documented during surgery (Figure 4) and the diagnosis of ICH caused by ICVT was confirmed. Postoperatively, the patient was admitted to our neurosurgical intensive care unit and therapeutic anticoagulation with UFH was reestablished. Right-sided weakness and aphasia improved gradually.

The patient was referred to rehabilitation 13 days after arriving at the hospital, with a Glasgow Coma Scale score of 15, diminished right hemiparesis, and reduced aphasia with a modified Rankin scale of 3.

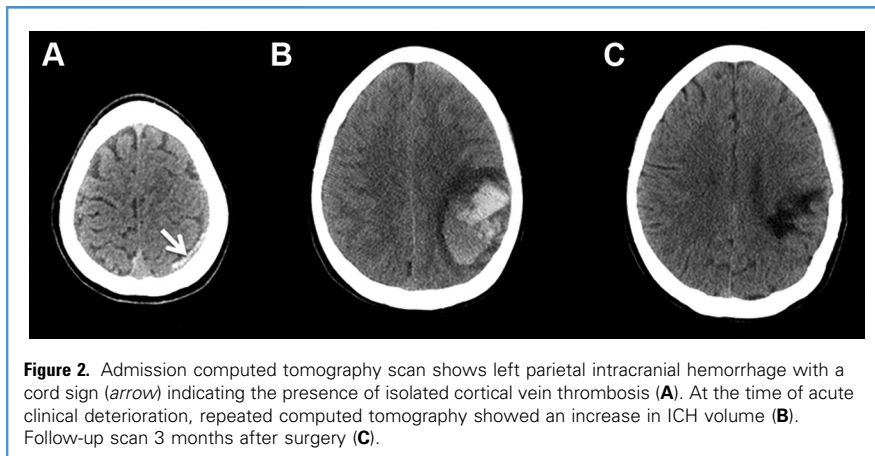
At 1 month follow-up, the patient presented with a complete recovery of symptoms (modified Rankin scale of 0). Blood analysis did not show any sign of coagulopathy or thrombophilia. The patient was treated with a vitamin K antagonist for 6 months.

### DISCUSSION

ICVT is a rare disease and clinical guidelines are scarce. Cortical vein thromboses are mostly seen in conjunction with SVT. A recent review published in *Stroke*<sup>2</sup> described 116 patients with ICVT with an average age of 41 years at time of presentation. ICVT symptoms were similar to those of sinus venous thrombosis, with the most common being headaches, followed by seizures and focal neurologic deficits.<sup>2</sup> It



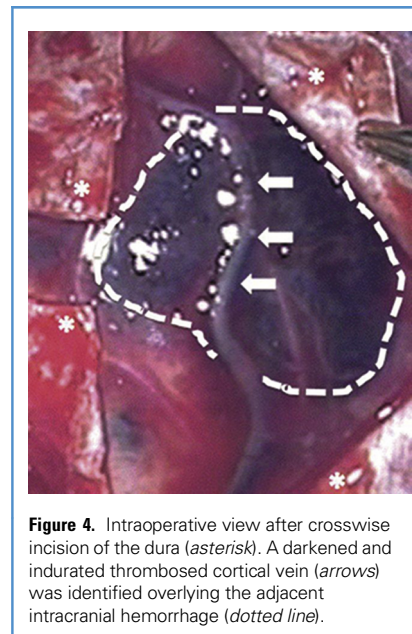
**Figure 1.** T2 fluid-attenuated inversion recovery magnetic resonance imaging examination 1 year before the event, showing no pathologic signs (A). At first appearance of symptoms and 4 days before intracranial hemorrhage magnetic resonance imaging T2 fluid-attenuated inversion recovery (B) and T1 contrast-enhanced sequences (C) revealed left-sided dural thickening (arrow) and enhancement (arrowhead).



**Figure 2.** Admission computed tomography scan shows left parietal intracranial hemorrhage with a cord sign (arrow) indicating the presence of isolated cortical vein thrombosis (A). At the time of acute clinical deterioration, repeated computed tomography showed an increase in ICH volume (B). Follow-up scan 3 months after surgery (C).

was reported that 16% of the patients had previously undergone dural puncture.<sup>2,9</sup> Pregnancy and puerperium are the most common risk factors because of hormonal changes and the associated prothrombotic effects.<sup>2,10</sup> The recommended approach for ICVT follows treatment guidelines for SVT.<sup>11</sup> From a pathophysiologic point of view, recanalization of the occluded vein is crucial in preventing the consequences of venous congestion (ie, brain edema and ICH). Consequently, even in the presence of ICH caused by venous congestion, therapeutic anticoagulation is indicated and fundamental.<sup>9,10</sup> A retrospective study of patients with SVT with associated ICH who were treated with intravenous heparin showed complete recovery in 52% of the cases. In patients with ICH who did not receive heparin, complete recovery was seen in only 23% of the patients, with a mortality of 69%.<sup>11,12</sup>

MRI is the standard tool for diagnosing ICVT<sup>3</sup> and may be followed by DSA to confirm the diagnosis.<sup>1</sup> Irregular signals detected on T2-weighted GE images within the thrombotic cortical vein (cord signs) are typical changes seen on MRI. Signal characteristics depend on the age of the thrombosis and any brain parenchymal signal abnormalities present (localized edema, hemorrhagic infarction, or ICH near the affected vein). Changes in brain parenchymal signal intensity are more common in ICVT than in SVTs.<sup>2</sup> Coutinho et al.<sup>2</sup> proposed that the high frequency of brain parenchymal lesions associated with ICVT, compared with sinus venous thrombosis, represented a possible selection bias, because case reports tend to describe severe cases more frequently. Signal loss in T2 and SWI-GE images has been found to be a highly sensitive sign.<sup>5,6</sup> In the present case, there were no SWI-GE sequences



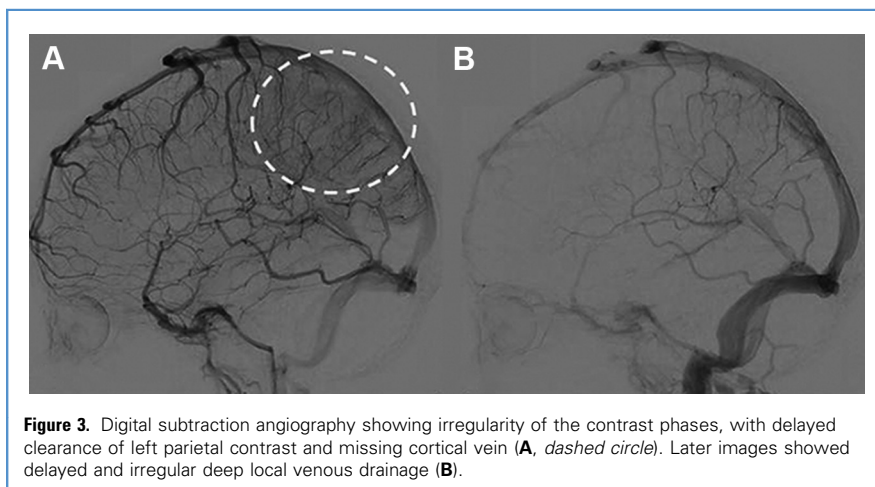
**Figure 4.** Intraoperative view after crosswise incision of the dura (asterisk). A darkened and indurated thrombosed cortical vein (arrows) was identified overlying the adjacent intracranial hemorrhage (dotted line).

available or any parenchymal signal changes on the initial MRI, only left hemispheric dural thickening and enhancement.

Cortical vein thrombosis may be easily identified as a cord sign on CT and MRI scans.<sup>3,4</sup> In our case, there was no cord sign on the first MRI, but only on the CT scan performed later for ICH. In patients without parenchymal lesions, diagnosing ICVT may be difficult, and those without a typical cord sign or neurologic symptoms could be missed, especially considering the average 7-day time span between the appearance of symptoms and diagnosis.<sup>2</sup> In our case, only ipsilateral dural thickening and enhancement in the left hemisphere were identified on the first MRI. Diffuse dural enhancement after dural puncture is a well-known phenomenon and is present in 83% of patients with CSF hypovolemia.<sup>7</sup> Differential diagnosis of diffuse dural enhancement includes neoplastic disorders (65%), CSF leakage, pachymeningitis, and sinus thrombosis.<sup>13</sup>

Incidental occurrence of asymptomatic cortical vein thrombosis on CT and MRI scans after LP has also been described (a cord sign in the affected vein and intraparenchymal lesions).<sup>8</sup> One pathophysiologic mechanism of an ICVT is suspected to be caused by the downward tearing of cortical veins caused by CSF hypovolemia.<sup>3</sup>

In our case, both the pregnancy and puerperium (which are the main risk



**Figure 3.** Digital subtraction angiography showing irregularity of the contrast phases, with delayed clearance of left parietal contrast and missing cortical vein (A, dashed circle). Later images showed delayed and irregular deep local venous drainage (B).

factors for ICVT, because of the hormonal changes),<sup>2</sup> as well as the EA potentially contributed to the onset of ICVT. The assumption that unilateral enhancement might be linked only to post-EA (with accidental dural puncture) is unlikely. We are not aware of any reports describing this phenomenon as a post-LP event.

DSA is recommended as a further diagnostic tool for inconclusive cases. ICVT can be diagnosed through indirect signs in the DSA such as collateral venous pathways, tortuous veins, delayed local venous drainage, or missing or rarefied contrast of the affected cortical vein.<sup>1</sup> In the DSA, our patient, who was suffering from ICH caused by ICVT, showed an irregularity of the contrast phases in the left parietal lobe with a missing cortical vein and a delayed and irregular regional venous drain.

To the best of our knowledge, ipsilateral dural thickening or enhancement after ICVT or after dural puncture (LP or by EA) has never been described in the literature. We propose that the thrombosis might already have been present at the time of the first MRI and that the ipsilateral left hemispheric dural enhancement was an indirect sign of an ICVT; possibly caused by blood congestion. Alternatively, if not evident on the first MRI, unilateral dural enhancement might identify patients at very high risk for imminent ICVT. Should this be confirmed by further cases/studies, it would warrant close monitoring and possibly early therapeutic anticoagulation. In the latter scenario, the cause of unilateral dural thickening must be elucidated. Because little is known about ICVT, many patients could be misdiagnosed or symptoms missed and (in contrast to the presented case) many patients show fewer neurologic symptoms and radiologic signs.<sup>2</sup> It is our opinion that the ipsilateral dural enhancement described in this case should be interpreted as an alarm signal

for possible ICVT and that further diagnostic testing would be required to rule it out. Once diagnosed, therapeutic anticoagulation with UFH or low-molecular-weight heparin should be started immediately to reduce the risk of complications such as ischemic infarction or ICH, which, even on its own, is associated with poor neurologic outcome.<sup>9</sup>

ICVT is a rare disease. We describe the first case of cortical vein thrombosis associated with ipsilateral dural thickening and enhancement shown on MRI, which potentially represents an indirect sign of ICVT.

Based on this case report, we should remain cautious and consider ICVT in patients presenting with ipsilateral dural enhancement, especially in combination with persistent position-independent headache. This may result in earlier treatment and anticoagulation therapy, potentially decreasing the risk of ICH and improving outcome.

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