Isolated Vein Thrombosis of the Posterior Fossa Presenting as Localized Cerebellar Venous Infarctions or Hemorrhages

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Background and Purpose—Cerebellar venous infarction or hemorrhage due to isolated venous thrombosis of the posterior fossa is a rare form of intracranial vein thrombosis that can be unsuspected in clinical practice.

Methods—We studied 230 patients with intracranial vein thrombosis, identifying 9 (3.9%; 7 women, mean age 34 years) with neuroimaging or histopathologic evidence of localized posterior fossa vein thrombosis causing parenchymal injury limited exclusively to the cerebellum.

Results—All patients had an insidious presentation suggesting other diagnoses. Intracranial hypertension (n=6) and cerebellar (n=4) syndromes were the main clinical presentations. Intracranial vein thrombosis was idopathic in 3 patients; associated with puerperium in 3; and with contraceptives, protein C deficiency, and dehydration in 1 case each. CT was abnormal but not diagnostic in 5 patients, showing a cerebellar hypodensity with fourth ventricle compression and variable hydrocephalus in 5 patients, and cerebellar hemorrhage in 2. Conventional MRI provided diagnosis in 6 cases, showing the causal thrombosis and cerebellar involvement; angiography was practiced in 2 of them, confirming the findings identified by MRI. In the other 3 patients, diagnosis was reached by histopathology. Thromboses were localized at the straight sinus (n=4), lateral sinuses (n=3), and superior petrosal vein (n=2). The acute case fatality rate was 22.2% (n=2), 1 (11.1%) patient was discharged in a vegetative state, 1 (11.1%) was severely disabled, and 5 (55.6%) were moderately disabled.

Conclusions—Isolated venous thrombosis of the posterior fossa is infrequent and implies a challenging diagnosis. Risk factors for intracranial vein thrombosis and atypical cerebellar findings on CT should lead to further MRI assessment.

Key Words: cerebellum ■ cerebral vein thrombosis ■ posterior fossa ■ sinus thrombosis ■ thrombosis

Intracranial vein thrombosis (IVT) is an infrequent condition that implies a wide spectrum of clinical manifestations and prognosis, ranging from mild headache to deep coma and from full recovery to death.1 The term isolated venous thrombosis of the posterior fossa is here used in reference to an infarction and/or hemorrhage resulting from localized thrombosis of the posterior venous drainage with parenchymal injury limited to the cerebellum. As shown by the medical literature, this is a rare form of IVT, and therefore, it is often unsuspected in clinical practice.2–8 As a consequence, scientific reports are scarce, mostly associating this condition with chronic suppurative processes or with surgical intervention of the posterior fossa. The parenchymal finding most frequently reported in isolated vein thrombosis of the posterior fossa is cerebellar venous infarction with or without a hemorrhagic component, but it can also present as a pure intracerebellar hemorrhage (Figure).2–8

We report on cases with isolated venous thrombosis of the posterior fossa selected from a large case series of Mexican patients with IVT. Our aim was to provide further knowledge on the clinical presentation, radiological features, and outcome at discharge of this rare form of venous thrombosis.

Methods

Nine cases of spontaneous isolated venous thrombosis of the posterior fossa were detected from a total of 230 consecutive patients with neuroimaging or histopathologic evidence of IVT admitted to 2 tertiary referral hospitals: Instituto Nacional de Neurología y Neurocirugía, Mexico City, Mexico.
cirugía “Manuel Velasco Suárez,” Mexico City (the first 200 cases, from 1973 to 1998), and Hospital Civil de Guadalajara, “Fray Antonio Alcalde” (the last 30 patients, from 1999 to 2008). The respective Committee of Ethics from both hospitals approved the study. All patients or their proxies provided informed consent. These patients were analyzed for clinical presentation, brain imaging, etiology, and outcome as assessed by the modified Glasgow Outcome Scale at discharge as follows: I=death; II=persistent vegetative state or total dependence for daily living; III=severe disability (conscious but disabled); IV=moderate disability (disabled but independent); and V=total recovery. We did not include secondary cases associated with suppurative processes or neurosurgical procedures of the posterior fossa. No venous MRI angiography or venous CT angiography was practiced on these patients, because this resource is of relatively recent introduction, and given the time in which this case series was started, the diagnostic workup was

Table. Clinical and Radiological Features of the Isolated Venous Thrombosis of the Posterior Fossa*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex/Age, Years</th>
<th>Clinical Presentation</th>
<th>Etiology</th>
<th>Diagnostic Resource Used</th>
<th>Initial Diagnostic Impression</th>
<th>Implicated Cerebellar Structures</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F/53</td>
<td>Nausea, dizziness, ataxia, IHS, stupor, and coma</td>
<td>Dehydration</td>
<td>Autopsy exclusively</td>
<td>Pan cerebellar syndrome</td>
<td>Vermian and right cerebellar hemisphere</td>
</tr>
<tr>
<td>2</td>
<td>F/42</td>
<td>Dizziness, ataxia, IHS, stupor, and coma</td>
<td>Not identified</td>
<td>CT/autopsy</td>
<td>Posterior fossa tumor</td>
<td>Left cerebellar hemisphere</td>
</tr>
<tr>
<td>3</td>
<td>M/56</td>
<td>Headache, ataxia, IHS, drowsiness</td>
<td>Not identified</td>
<td>CT/MRI</td>
<td>Posterior fossa tumor</td>
<td>Left cerebellar hemisphere</td>
</tr>
<tr>
<td>4</td>
<td>F/16</td>
<td>Dizziness, ataxia, drowsiness</td>
<td>Puerperium</td>
<td>CT/MRI</td>
<td>Hemispheric cerebellar and vermian infarction</td>
<td>Right cerebellar hemisphere</td>
</tr>
<tr>
<td>5</td>
<td>F/25</td>
<td>Ataxia, IHS</td>
<td>Puerperium</td>
<td>CT/MRI</td>
<td>Pan cerebellar infarction</td>
<td>Vermian and right cerebellar hemisphere</td>
</tr>
<tr>
<td>6</td>
<td>F/18</td>
<td>Dizziness, ataxia</td>
<td>Puerperium</td>
<td>CT/MRI/4-vessel angiography</td>
<td>Cerebellar hemorrhagic infarction</td>
<td>Right cerebellar hemisphere</td>
</tr>
<tr>
<td>7</td>
<td>F/33</td>
<td>Dizziness, ataxia, drowsiness</td>
<td>Oral contraceptives</td>
<td>CT/MRI</td>
<td>Hemispheric cerebellar infarction</td>
<td>Right cerebellar hemisphere</td>
</tr>
<tr>
<td>8</td>
<td>M/14</td>
<td>Dizziness, ataxia, IHS, drowsiness</td>
<td>Protein C deficiency</td>
<td>CT/biopsy</td>
<td>Cerebellar tumor</td>
<td>Left cerebellar hemisphere</td>
</tr>
<tr>
<td>9</td>
<td>F/63</td>
<td>Dizziness, IHS, drowsiness</td>
<td>Not identified</td>
<td>CT/MRI/4-vessel angiography</td>
<td>Cerebellar hemorrhage</td>
<td>Left cerebellar hemisphere</td>
</tr>
</tbody>
</table>

*The order in this table reflects that of the clinical identification of each case. F indicates female; M, male.
heterogeneous, which includes thrombophilia investigation. Hence, this communication mainly focuses on clinical and neuroimaging findings. Final diagnosis was achieved by means of autopsy (n=2), brain MRI (n=6), and cerebellar biopsy (n=1). We obtained standard MRI techniques current to the time in every patient was seen, mainly 0.5- to 1.5-T MRI in T1, T2, and fluid-attenuated inversion recovery (only in the last 2 patients) sequences. Gradient echo/T2* sequences could not be obtained for any patient. Involvement of cerebellar veins was identified by histopathologic and neuroimaging (when possible) analyses. Four-vessel angiography was practiced on 2 patients, 1 of them with cerebellar veins involvement. Autopsy allowed for a fine determination of the cerebellar veins implicated (n=2; Table). Neuroimaging techniques permitted only gross inferences with respect to the cerebellar veins involved; therefore, for homogeneity here the term “cerebellar veins” is mentioned without a precise depiction of each of them. Descriptive statistics are presented as simple frequencies and percentages. For analyses on outcome, relative frequencies are calculated with the respective 95% CIs by the Wald method. SPSS Version 13.0 (Chicago, Ill) was used for statistical calculations.

Results
From a total of 230 patients, 9 (3.9%) were diagnosed with isolated venous thrombosis of the posterior fossa (7 women, mean age 34 years, range 14 to 63 years). All patients had a subacute presentation characterized by an insidious installation of neurological features in >48 hour but in <30 days. All cases presented clinically suggesting other diagnoses: 4 patients had cerebellovestibular symptoms before hospital presentation and 5 developed intracranial hypertension syndrome. IVT was associated with puerperium in 3 cases and with contraceptives, protein C deficiency, and dehydration in 1 case each (Table). No obvious cause or risk factor was identified in 3 patients. Brain CT was practiced to 8 cases, being abnormal in all, but without suggesting the specific diagnosis (CT showed cerebellar hypodensities, pseudotumoral mass effect, hemorrhage, and variable degree hydrocephalus). MRI was abnormal in the 6 patients who received this assessment, showing the sinovenous thrombosis and cerebellar involvement in all. Straight sinus (n=4), left lateral sinus (n=2), right lateral sinus (n=1), and superior petrosal vein (n=2) were the venous systems affected. Two patients died in the acute state due to intracranial hypertension syndrome (IHS) and had autopsies that provided definite evidence of isolated venous thrombosis of the posterior fossa. Three patients received anticoagulants, 2 patients received antiplatelets, and 1 had steroids for treatment. Suboccipital decompression with a wide biopsy was performed in 1 patient (Case 8, Table) due to an initial suspicion of posterior fossa tumor. None of the remaining patients received a shunt or decompressive surgery. Acute case fatality rate was 22.2% (n=2; 95% CI: 5.3% to 55.7%), 1 (11.1%, 95% CI: 0.001% to 45.7%) patient was discharged in a vegetative state, 1 (11.1%, 95% CI: 0.001% to 45.7%) was severely disabled, and 5 (55.6%, 95% CI: 26.6% to 81.2%) were moderately disabled (Table). At hospital discharge, no cases with complete recovery were observed.

Discussion
The low frequency of isolated venous thrombosis of the posterior fossa in our case series reveals the rarity of this form of IVT and is in accordance with the largest prospective collaborative multicenter international study of cerebral venous thrombosis (International Study on Cerebral Vein and Dural Sinus Thrombosis [ISCVT], n=624), in which venous infarction of the posterior fossa was reported by CT/MRI in 3.2% and parenchymal hemorrhage in 1.6%. However, from the primary data provided in that report, we could not exclude the simultaneous extension of the thrombosis into the cerebral superficial or deep sinuses as well as the possible implication of supratentorial structures; thus, further comparisons with our cases are not possible. Of the remaining 221 patients of our case series, no information could be obtained on how many of them had simultaneous implication of supra- and infratentorial structures or veins, which could provide useful information on diagnosis and outcome, in comparison with our 9 patients here reported with implication limited to the infratentorial region.

Venous infarctions in the posterior fossa result from thrombosis of the lateral and straight sinuses as well as the superior petrosal vein. In our present report, the most frequent sinuses affected were the straight and lateral followed by the petrosal vein. The reason for the rarity of isolated venous thrombosis of the posterior fossa could be the abundant collateral venous drainage of the posterior structures, which prevents blood flow stasis in this area. A thrombosis of lateral or straight sinuses usually implies lesions in supratentorial parenchyma. A recent single-center
analysis on 62 cases with isolated lateral sinus thrombosis did not report cases with cerebellar infarction or hemorrhage, and most parenchymal abnormalities were confined to the supratentorial structures.10

The clinical importance of the isolated venous thrombosis of the posterior fossa is the difficulty in making the diagnosis based on the initial clinical and neuroimaging findings. Clinicians should be aware of this differential diagnosis in a particular patient who has risk factors for IVT presenting with cerebellovestibular symptoms, headache, intracranial hypertension syndrome, and atypical findings of the posterior fossa structures on brain CT (ie, pan cerebellar and vermic infarcts, cerebellar hemorrhages of irregular shapes, or with extension to the subarachnoid space and cerebellar peduncles). A brain CT should be the initial diagnostic resource that would prompt MRI assessment in cases highly suggestive of isolated vein thrombosis of the posterior fossa. In ideal grounds, a venography (either venous CT or MRI) should be confirmatory. This form of IVT is a differential diagnosis of presumptive rapidly growing cerebellar neoplasms, because they can also have an acute or subacute presentation with a mass effect, perilesional edema, intratumoral bleeding, and compression of the fourth ventricle. Furthermore, it has been reported that a venous infarction due to isolated venous thrombosis of the posterior fossa can also present gadolinium enhancement, making the diagnostic analysis even more confusing.8

Indeed, our study has several limitations that should be addressed. This is a retrospective analysis of patients prospectively included in a research database designed to address different objectives. Also, the follow-up period is limited to hospital discharge, because further information was lost for most patients, which includes the rest of the IVT cases, whose clinical comparison with the case series here reported would be useful to emphasize meaningful differences.

In conclusion, a clinician should investigate the possibility of isolated venous thrombosis of the posterior fossa in the presence of known risk factors and atypical posterior fossa lesions on neuroimaging. The prognosis of this type of IVT may imply a higher frequency of unfavorable outcomes as compared with other IVT forms, an issue that should be investigated in comparative analyses. Prompt recognition of this entity is essential for adequate management.

Disclosures

None.

References

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