Subarachnoid neurocysticercosis and an intracranial infectious aneurysm: case report

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Infectious intracranial aneurysms (IIAs) represent 2%–6% of all intracranial aneurysms and, classically, have been associated with bacterial or fungal agents. The authors report the case of a 42-year-old woman who presented with a typical history of subarachnoid hemorrhage. Digital subtraction angiography (DSA) showed an aneurysmal dilatation on the frontal M2 segment of the left middle cerebral artery (MCA). The patient was treated surgically, and multiple cysts were found in the left carotid and sylvian cisterns, associated with a dense inflammatory exudate that involved the MCA. The cysts were removed, and a fusiform aneurysmal dilatation was identified. The lesion was not amenable to direct clipping, so the authors wrapped it. Histopathological analysis of the removed cysts revealed the typical pattern of subarachnoid neurocysticercosis. The patient received cysticidal therapy with albendazole and corticosteroids, and she recovered uneventfully. Follow-up DSA performed 6 months after surgery showed complete resolution of the aneurysm. The authors performed a review of the literature and believe that there is sufficient evidence to affirm that the subarachnoid form of neurocysticercosis may lead to the development of an IIA and that Taenia solium should be listed among the possible etiological agents of IIAs, along with bacterial and fungal agents.

https://thejns.org/doi/abs/10.3171/2019.5.FOCUS19280

KEYWORDS subarachnoid hemorrhage; infectious intracranial aneurysm; neurocysticercosis; stroke
surface of the membrane with calcified foci (Fig. 3). No scolex was identified, which is typical in SNCC. After surgery, fundoscopy was performed, and ocular cysticercosis was ruled out. The patient then received cysticidal therapy with albendazole (15 mg/kg/day for 7 days) and corticosteroids (prednisone 1 mg/kg/day for 3 weeks, starting 3 days prior to the administration of albendazole) and recovered uneventfully. Follow-up DSA, performed 6 months after surgery, showed resolution of the aneurysm and complete regression of irregularities on the vessel caliber (Fig. 4). Forty-eight months after hospital discharge, the patient remains asymptomatic and has fully returned to her routine daily activities.

Discussion

SNCC occurs less frequently than the parenchymal form, but it is much more severe. The *T. solium* larvae can reach the subarachnoid space through the choroid plexus, traveling throughout the ventricular system, and finally reaching the basal cisterns, or directly through the meningeal vessels. In SNCC, these larvae usually develop into the racemose form of neurocysticercosis, which differs from the cellulosae form found in the parenchymal variant of neurocysticercosis. In the racemose form, the cysts tend to grow to larger diameters than in the cellulosae form (50 vs 10 mm on average), grouping in clusters that resemble bunches of grapes. It appears that the larger volume of the cysts in the racemose form is due to the greater existing space in the basal cisterns and to the inflow of CSF into the cyst, contributing to the inability to visualize the scolex in this variant of the disease. A hallmark of SNCC and the racemose form is the strong inflammatory reaction caused by the presence of cysts in the subarachnoid space, leading to a dense exudate composed of lymphocytes, eosinophils, giant cells, collagen fibers, and hyalized parasitic membranes, as well as high CSF levels of cytokines interleukin-1, -5, and -6 and tumor necrosis factor–α, which are probably secreted by peripheral lymphocytes and activated microglia. Such inflammatory reaction, resulting in chronic arachnoiditis, seems to be the key to understanding the clinical events related to SNCC.

Patients with SNCC most often present with hydroceph-
alus as well as signs of intracranial hypertension, which occurs due to obstruction of the basal CSF pathways and arachnoid granulations. This obstruction is caused both by the presence of the cysticerci and the associated inflammatory reaction. Another possible presentation is cranial nerve neuropathy, caused by inflammatory arachnoiditis and/or direct compression by a racemose cyst.7

Vascular events are unusual manifestations of SNCC and occur in only 3%–12% of cases of NCC.1,10 In most cases, these events are ischemic strokes involving perforating arteries and leading to lacunar infarcts, but they may also affect larger arteries, such as the MCA, and lead to extensive infarctions.16,20,21 Hemorrhagic presentation is rare, with few reports in the literature. Inflammation and chronic arachnoiditis affecting the artery wall may lead to occlusive endarteritis, causing ischemic strokes, or weakening of the vessel wall and the formation of aneurysms and bleeding.24 In a study by Barinagarrementeria and Cantú, patients with SNCC underwent DSA that showed radiological signs of arteritis in 58% of cases, which more frequently involved the MCAs and posterior cerebral arteries.7

Infectious Intracranial Aneurysms and SNCC

In 1869, Church was the first to report the occurrence of an IIA in a 13-year-old boy with bacterial endocarditis.6 Osler coined the term “mycotic aneurysm” when he described an aortic aneurysm in a patient with bacterial endocarditis, due to the appearance of “fresh fungus vegetations.”19 It is known, however, that most mycotic aneurysms are actually of bacterial origin. A change in the nomenclature was then proposed and the term “infectious (or infected) aneurysm” replaced mycotic aneurysm.3

IIAs represent approximately 2%–6% of all intracranial aneurysms, and, in children, it may represent as many as 10%.15,26 The etiological agent in most cases is bacteria. A fungal etiology is commonly associated with congenital or acquired immunodeficiency states. The pathophysiological mechanism of IIA formation may be by intravascular or extravascular spread. Intravascular spread occurs through septic emboli that are commonly of cardiac origin. In such cases, the infectious aneurysm tends to occur more distally, usually on the MCA. In contrast, IIAs formed by extravascular spread result from vessel-wall injury and weakening due to the extension of contiguous infectious foci; they usually affect larger-caliber arteries of the cranial base and lead to the formation of more proximal aneurysms compared to those of intravascular etiology. IIAs of bacterial etiology are usually formed by intravascular spread, whereas those of fungal etiology tend to form from extravascular mechanisms.18

To the best of our knowledge, there are 8 case reports in the medical literature describing the occurrence of SAH secondary to aneurysms related to SNCC (Table 1). In all of the cases in which surgery was performed, a dense inflammatory reaction/infiltrate was noted along with the
presence of many cysticerci surrounding and adhered to the involved vessel, which, in most cases, was the MCA. In addition, the aneurysm was found to be friable and non-saccular and, in most cases, was treated with alternative methods to direct clipping (i.e., trapping or wrapping). Kim et al. were the only authors to perform histological analysis of the aneurysm wall; the analysis showed diffuse lymphocytic infiltration and thinning of the vessel wall with no internal elastic lamina, which are common findings in IIAs. Marquez-Romero et al. and Cárdenas et al. reported on the conservative treatment of an MCA aneurysm with albendazole and prednisone with good clinical and radiological results, what has also been demonstrated for IIA of other etiologies. We, therefore, believe that there is sufficient evidence that NCC, in its subarachnoid form, through an extravascular inflammatory mechanism affecting the wall of the involved vessel, may lead to the development of an IIA and should be listed among the possible etiological agents, along with bacterial and fungal agents.  

There should be a high level of suspicion for IIAs due to SNCC in patients who have parenchymal lesions, but it will be extremely difficult to diagnose in patients who do not have those lesions. The results of CSF analysis may be normal, including negative antigen and antibody tests. Furthermore, intracranial hemorrhage and/or hydrocephalus may make lumbar puncture contraindicated. Radiological diagnosis is also difficult to establish because, in the racemose form, the parasites have a signal intensity identical to that of the CSF, do not show contrast enhancement, and do not have any scoleces. Often, subtle changes such as a unilateral enlargement of a basal cistern will be the only diagnostic clue. The acquisition of FLAIR sequences after 5 minutes of continuous inhalation of 100% supplemental oxygen may have a higher sensitivity for cysticercus visualization.  

Although there is report of success with the conservative treatment with albendazole at high doses (30 mg/kg/day) and corticosteroids (intravenous dexamethasone 24 mg/day, followed by oral prednisone 1 mg/kg/day), we do not recommend such an approach. Unlike antibiotic treatment for IIAs from a bacterial or fungal origin, there is doubt about the efficacy of cysticidal drugs for the treatment of SNCC. In most cases, an aneurysm-securing treatment should be performed. In our review, surgery was performed in 6 of the 8 cases, and we believe this should be the preferred method of treatment because it allows the mechanical removal of the cysts, reducing the inflammatory reaction and also the obstruction of CSF pathways through the basal cisterns. The method of aneurysm treatment during surgery should be evaluated on a case-by-case basis. In only 1 reported case was conventional clipping possible. In the other cases, the aneurysm was treated by trapping or wrapping in conjunction with cysticidal treatment. Depending on the aneurysm location, trapping and extracranial-intracranial or intracranial-intracranial bypass may be an option. After surgical treatment, cysticidal drugs should be administered in all cases. For our patient, as mechanical removal of the cysts was performed, a dosage of 15 mg/kg/day of albendazole was administered for 7 days along with oral prednisone (1 mg/kg/day) for 21 days, starting 3 days prior to albendazole administration. It is important to emphasize that, in all reports, including ours, the outcomes were good, with clinical improvement, which implies, perhaps, a better prognosis of the IIA.

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Presentation</th>
<th>Findings</th>
<th>Treatment</th>
<th>HS Findings</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Zee et al., 1980</td>
<td>23, M</td>
<td>Temporal hematoma</td>
<td>Distal MCA aneurysm</td>
<td>NA</td>
<td>Proximal artery clipping</td>
<td>Cysticercus</td>
</tr>
<tr>
<td>Soto-Hernandez et al., 1996</td>
<td>32, M</td>
<td>SAH</td>
<td>M1 MCA aneurysm</td>
<td>Cells; protein; glucose + ELISA for NCC</td>
<td>Wrapping</td>
<td>Cysticercus</td>
</tr>
<tr>
<td>Huang et al., 2000</td>
<td>32, M</td>
<td>SAH</td>
<td>Proximal MCA aneurysm</td>
<td>NA</td>
<td>Clipping</td>
<td>Cysticercus</td>
</tr>
<tr>
<td>Kim et al., 2005</td>
<td>69, M</td>
<td>SAH</td>
<td>Proximal MCA aneurysm</td>
<td>NA</td>
<td>Trapping</td>
<td>Cysticercus &amp; aneurysm wall</td>
</tr>
<tr>
<td>Marquez-Romero et al., 2012</td>
<td>38, M</td>
<td>SAH</td>
<td>M1 MCA aneurysm</td>
<td>Normal</td>
<td>Conservative (albendazole &amp; steroids)</td>
<td>NA</td>
</tr>
<tr>
<td>Cárdenas et al., 2012</td>
<td>39, F</td>
<td>SAH</td>
<td>M1 MCA aneurysm</td>
<td>Cysts in sylvian fissure</td>
<td>Wrapping</td>
<td>Cysticercus</td>
</tr>
<tr>
<td>33, M</td>
<td>SAH</td>
<td>Distal MCA aneurysm</td>
<td>NA</td>
<td>Cysts in both sylvian fissures</td>
<td>Conservative (albendazole &amp; steroids)</td>
<td>Cysticercus</td>
</tr>
<tr>
<td>Eboli et al., 2012</td>
<td>80, M</td>
<td>SAH</td>
<td>Distal MCA aneurysm</td>
<td>NA</td>
<td>Clipping</td>
<td>Cysticercus</td>
</tr>
<tr>
<td>Present case</td>
<td>42, F</td>
<td>SAH</td>
<td>M1 MCA aneurysm</td>
<td>NA</td>
<td>Wrapping</td>
<td>Cysticercus</td>
</tr>
</tbody>
</table>

AICA = anterior inferior cerebellar artery; ELISA = enzyme-linked immunosorbent assay; HS = histopathological study; NA = not available.
related to SNCC when compared to those of bacterial or fungal origin.

Conclusions

NCC may present with many clinical manifestations, and, in its subarachnoid form, can lead to the formation of IIA through an extravascular inflammatory mechanism. The possibility of an IIA due to SNCC should always be considered when a patient with SAH has associated parenchymal lesions or asymmetrical enlargement of a basal cistern. In our opinion, the treatment of choice is microsurgery, as it allows occlusion of the aneurysm and mechanical removal of racemose cysts, followed by standard drug treatment. Although there are reports of success with the conservative treatment with cysticidal drugs and corticosteroid agents alone, this alternative should be reserved for selected cases associated with high surgical risks and not amenable to endovascular treatment, because there are still doubts about the effectiveness of this type of treatment in SNCC. An SAH due to IIA associated with SNCC seems to have a better prognosis compared with IIA of bacterial or fungal origin.

References


Disclosures

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions

Conception and design: Vieira, Faquini, Cezar. Acquisition of data: Vieira, Silva, Cezar. Analysis and interpretation of data: Vieira, Silva, Griz, Cezar. Drafting the article: Vieira, Silva, Griz. Critical revision of the article: Vieira, Faquini, Silva, Griz, Almeida, Azevedo-Filho. Reviewed submitted version of manuscript: Vieira, Almeida, Azevedo-Filho. Approved the final version of the manuscript on behalf of all authors: Vieira. Statistical analysis: Azevedo-Filho. Study supervision: Vieira.

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