Massive Cerebellopontine Angle Cysticercosis: current treatment aspects

Cisticercose do Ângulo Pontocerebelar: aspectos atuais do tratamento

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ABSTRACT

Background: neurocysticercosis is the most common Central Nervous System infection and is caused by Taenia solium. The symptoms vary depending on the lesion’s localization in the brain. The cerebellopontine angle is a rare location of cysticercosis. Only 14 cases were reported.

Case presentation: a 50-year-old man presented headache and left facial trigeminal neuralgia. He reported a previous history of hydrocephalus. Glasgow coma scale was 14. There was left facial hypoesthesia and left peripherical facial palsy. MRI showed multiple cystic lesions in the extra-axial space localized on the left cerebellopontine angle and the right Sylvian fissure. The patient was submitted for surgical treatment.

Conclusion: cerebellopontine angle neurocysticercosis is a rare entity that determines severe symptoms and the risk of long-time morbidity and mortality of patients. Only 15 cases were reported in the Literature and different treatment options exist, including ours.

Keywords: Neurocysticercosis; Cerebellopontine angle; Neurosurgery; CNS infection

RESUMO


Palavras-chave: Neurocisticercose; Ângulo pontocerebelar; Neurocirurgia; Infecção do SNC
INTRODUCTION

Neurocysticercosis is a Central Nervous System infection caused by Taenia solium. The first description report was in 1946 and, since then, turned into the most common CNS infection worldly1-3.

The symptoms vary depending on the lesion's localization in the brain. The most common are headaches and seizures. However, extra-parenchymal cysticercosis could be very distinct clinically compared to the parenchymal form3.

The subarachnoid space is the most common extra-parenchymal form and could lead to CSF block and surrounding inflammation, including cranial nerve dysfunction. In these conditions, cysticercus occasionally develops clusters and could form racemose cysts3-7.

The cerebellopontine angle is a rare and uncommon location of neurocysticercosis. The first description was in 1992. Since then, only 14 cases were reported2,4-6,8-17, and 15 if included ours.

CASE PRESENTATION

A 50-year-old man was admitted to the emergency department of our facilities presenting severe headache and left facial trigeminal neuralgia which started 15 days before admission. He had a previous history of obstructive hydrocephalus treated with a ventriculoperitoneal shunt in 2020 in another facility.

On neurological examination, the patient was confused, and the Glasgow coma scale was 14. There were left facial hypoesthesia and left peripheral facial palsy House & Brackmann grade 3. A bilateral paresis worse on the left side was observed. Because of these symptoms, an MRI was done and showed multiple cystic lesions in the extra-axial space localized on the left cerebellopontine angle and the right Sylvian fissure (Figure 1).

During the time of evaluation, the patient presented a worse neurological function with somnolence and dysphagia with paresis of the left caudal nerves. Because of this and localizing the main symptoms, it was decided to have him operate on the left cerebellopontine angle lesion.

In surgery, the appearance was consistent with multiple cystic lesions, and several scolexes were identified (Figure 2). After the lesions removal, the brainstem was decompressed, and all the cerebellopontine cistern was free. The pathologic study confirmed cysticercosis.

After the surgical treatment, the patient presented improvement in the headache and trigeminal facial neuralgia. No changes were achieved in motor function and facial paresis, however.

DISCUSSION

The first description of neurocysticercosis dates from 1946 by Albo. Since then, this diagnosis became the most common Central Nervous System parasite infection, and several treatments and approaches were discussed1,3,18.

Almost as accident, this pathology results from ingesting water and/or food contaminated with eggs of Taenia solium. This is the wrong cycle of life of the pathogen which affects commonly pigs and humans. During this atypical lifecycle, after the ingestion of eggs, in the intestinal tract, the release of the egg-enclosed larvae perforates the wall of the intestine and is transported to several tissues like the brain3,7.

Despite the actual knowledge, neurocysticercosis remains a neglected tropical disease. The World Health Organization estimates around 2.5 to 8 million people have with this diagnosis worldly. Asia (including India, China, and Nepal), Latin America (including Brazil), and Africa are the most common areas3,7,8.

Despite a common disease, the clinical and morphological presentations are different around the world. For example, in India it is more frequently a single and isolated parenchymal form, while in Latin America the most common is multiple cysticerci in extra-parenchymal form3-7.
Figure 1. A-D. Preoperative MRI.

Figure 2. A-B. Surgical aspect.
There are two forms of Neurocysticercosis: parenchymal and extra-parenchymal. These morphological aspects affect the clinical presentation and the treatment of pathology. The first form is more common. The cysticerci are deposited inside the grey and white junction. Headache and seizures are the main symptoms, nevertheless, motor, psychiatric, and language symptoms are described\(^3,7,9,11,12,18\).

The extra-parenchymal form is heterogeneous. In general, this form presents a poor prognosis with higher morbidity and mortality. The correct treatment remains unclear. Because of the dissemination inside the subarachnoid space, severe inflammation is often started, and the patients could develop hydrocephalus, high intracranial pressure, vasculitis, and cranial nerve dysfunction. Another potential complication is the aggregation of cysts forming the racemose cyst\(^3,7,19\).

Several diagnosis methods are described in the Literature. Immunological analysis, microscopy, and neuroimage represent the modalities used for diagnostic criteria. In this classification\(^3,7\), the absolute criteria include histology, fundoscopy, or scolex visible within the cystic lesion in MRI\(^3,7\). Other possibilities variables include major and minor criteria as a positive result for the detection of T. solium antibodies, clinical manifestations, positive CSF Elisa, and lesion resolution after cytocidal drug therapy.

Because of the importance of diagnosis, the MRI is indispensable. The main characteristics are the presence of scolex, cysts, racemous form, and calcifications. Other possible alterations are perilesional edema and the starry sky (that is formed by the presence of multiple cysts during different stages)\(^3,7,18\).

The treatment depends on the lesion’s localization and patient status. There are many options as the surgical treatment that is indicated for massive racemose form and some subarachnoidal forms. The use of cysticidal drugs is an important cluster of the treatment, however, patients with a risk of hydrocephalus and subarachnoidal form represent a contraindication for the use. Steroids also are essential. These drugs regulate inflammation and reduce the risk of potential complications\(^2-4,7,10,13,14,17-19\).

Despite being described as possible, only a few cases of cerebellopontine angle cysticercosis were related (Table 1)\(^2,4-6,8-17\). Most of the cases were treated with surgical excision of the lesions (93.3%). Only 1 case was treated with a cysticidal drug alone.

The most common symptoms in cerebellopontine angle neurocysticercosis are hearing loss, headache, and trigeminal neuralgia. The surgical treatment was effective and showed a faster improvement in the patients compared with the unique patient treated clinically.

### Table 1. Described cerebellopontine angle cysticercosis.

<table>
<thead>
<tr>
<th>Case</th>
<th>Author, Year</th>
<th>Age, Sex</th>
<th>Presentation</th>
<th>Management</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Celis et al., 1992(^{10})</td>
<td>36yo. Woman.</td>
<td>Anacusis and imbalance</td>
<td>Surgical Treatment</td>
<td>Improvement of symptoms</td>
</tr>
<tr>
<td>2</td>
<td>Revuelta et al., 1995(^{14})</td>
<td>52yo. Man.</td>
<td>Trigeminal neuralgia</td>
<td>Surgical Treatment</td>
<td>Improvement of symptoms</td>
</tr>
<tr>
<td>3</td>
<td>del Brutto, 2000(^{11})</td>
<td>28yo. Woman.</td>
<td>Anacusis and contralateral hemiparesis</td>
<td>Praziquantel</td>
<td>Improvement of symptoms</td>
</tr>
<tr>
<td>4</td>
<td>Aguiar et al., 2000(^{8})</td>
<td>42yo. Woman.</td>
<td>Bilateral trigeminal neuralgia</td>
<td>Surgical Treatment</td>
<td>Improvement of symptoms</td>
</tr>
<tr>
<td>5</td>
<td>Revuelta et al., 2003(^{6})</td>
<td>59yo. Woman.</td>
<td>Trigeminal neuralgia</td>
<td>Surgical Treatment</td>
<td>Improvement of symptoms</td>
</tr>
<tr>
<td>6</td>
<td>Chang et al., 2004(^{9})</td>
<td>60yo. Man.</td>
<td>Anacusis and imbalance</td>
<td>Surgical Treatment and Praziquantel</td>
<td>Improvement of symptoms</td>
</tr>
<tr>
<td>7</td>
<td>Jarupant et al., 2004(^{12})</td>
<td>52yo. Woman.</td>
<td>Bilateral CPA symptoms</td>
<td>Surgical Treatment and albendazole</td>
<td>Death</td>
</tr>
<tr>
<td>8</td>
<td>Singh et al., 1999(^{16})</td>
<td>39yo. Man.</td>
<td>Imbalance and headache</td>
<td>Surgical Treatment</td>
<td>Not described</td>
</tr>
</tbody>
</table>

CPA: Cerebellopontine Angle
CONCLUSION

Cerebellopontine angle neurocysticercosis is a rare entity that determines severe symptoms and the risk of long-time morbidity and mortality of patients. Only 15 cases were reported in the Literature and there are different treatment options, including our case.

A better treatment condition – if exist brainstem compression and neurological symptoms – is offered to the patient for a retrosigmoid craniotomy and excision of the cysticercosis. The use of steroids is an important key to reducing the risk of arachnoid inflammation and the morbidity associated with pathology.

REFERENCES


Table 1. Continued...

<table>
<thead>
<tr>
<th>Case</th>
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<th>Presentation</th>
<th>Management</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>9</td>
<td>Song et al., 2013</td>
<td>68yo. Woman.</td>
<td>Hearing loss</td>
<td>Surgical Treatment and Praziquantel</td>
<td>Improvement of symptoms</td>
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<td>10</td>
<td>Karegowda et al., 2014</td>
<td>43yo. Man.</td>
<td>Anacusis and imbalance</td>
<td>Surgical Treatment and Praziquantel</td>
<td>Not described</td>
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<td>11</td>
<td>Kumar et al., 2018</td>
<td>58yo. Woman.</td>
<td>Anacusis and imbalance</td>
<td>Surgical Treatment</td>
<td>Improvement of symptoms</td>
</tr>
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<td>12</td>
<td>Pandita and Razdan, 2019</td>
<td>30yo. Man.</td>
<td>Headache</td>
<td>Steroids and Albenzadol</td>
<td>Improvement of symptoms</td>
</tr>
<tr>
<td>13</td>
<td>Yang and Vásquez, 2020</td>
<td>30yo. Man.</td>
<td>Headache</td>
<td>Surgical Treatment</td>
<td>Improvement of symptoms</td>
</tr>
<tr>
<td>14</td>
<td>Choudhary et al., 2021</td>
<td>32yo. Man.</td>
<td>Anacusis and hemiparesis.</td>
<td>Surgical Treatment, Steroids and albendazol</td>
<td>Presented improvement of paresis</td>
</tr>
<tr>
<td>15</td>
<td>This case, 2023</td>
<td>50yo. Man.</td>
<td>Headache, trigeminal neuralgia, and hemiparesis</td>
<td>Surgical Treatment, Steroids and albendazol</td>
<td>Presented improvement of headache and trigeminal neuralgia</td>
</tr>
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</table>

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