Have you washed your hands today? A review of neurocysticercosis.

Poster No.: C-1033
Congress: ECR 2015
Type: Educational Exhibit
Authors: L. G. Campos, J. Zampieri, F. D. S. Souza, M. G. Longo, J. Á. Duarte, J. A. Perez, L. Vedolin; Porto Alegre/BR
Keywords: MR, CT, Neuroradiology brain, Education, Infection
DOI: 10.1594/ecr2015/C-1033

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR's endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method is strictly prohibited.
You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.
Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

- Review the epidemiology, transmission and pathogenesis of cysticercosis

- Describe the most common clinical features and imaging findings

- Correlate the stages of infection with imaging findings in the central nervous system (CNS)
Background

1. Terminology and Etiology

Neurocysticercosis is the most common helminthic infection of the nervous system caused by the encysted larva of the tapeworm *Taenia solium*. It is the most important parasitic disease of the human CNS and a leading cause of epilepsy in underdeveloped world (1). CNS lesions eventually develop in 60-90% of patients with cysticercosis (2). Neurocysticercosis refers specifically to CNS involvement by cysticercosis. Although involvement of muscle and subcutaneous tissues by this disease is frequently asymptomatic, within the CNS cysticerci can produce a variety of clinical neurologic symptoms (3).

The complex life cycle of *Taenia solium* involves two hosts (Fig. 1). Humans are the only definitive hosts for the adult tapeworm, whereas both pigs and humans may act as intermediate hosts for the larval form called cysticercus. In the normal cycle of transmission, the adult *T. solium* inhabits the small intestine of humans (typically from ingestion of uncooked pork), where it is attached to the intestinal wall by its potent suckers and hooks (4). The intestinal infection by this parasite is referred to as taeniasis, which in many cases is asymptomatic; however, it may occasionally cause mild diarrhea and abdominal pain. Consequently many individuals with taeniasis are not aware of their disease. The pig (intermediate host) typically acquires the parasite by injection of feed contaminated with human fecal material containing *T solium* eggs (oncospheres) (3). The larval forms of the parasite emerge from the eggs in the gastrointestinal tract, penetrate the wall of the bowel, and disseminate via the bloodstream throughout the muscle tissue of the pig, forming cysts in the surrounding tissue, known as cysticerci (3).

Humans also can develop cysticercosis when it is acquired through the ingestion of eggs from the feces of a tapeworm carrier, with little evidence of other forms of contamination (eg, through the agency of air, water, or flies) (1). Human cysticercosis infection does not necessarily require contact with infested animals, only with a tapeworm carrier (1). When humans ingest food contaminated with human feces containing *T solium* eggs, the larvae similarly invade the bloodstream of the humans and disseminate throughout the tissues of the body, forming cysticerci at terminal arterioles within the CNS, subcutaneous tissues, muscles, and so forth (3).

2. Epidemiology
Clinically, the magnitude of the disease burden associated with neurocysticercosis in endemic countries is enormous; it is the most common cause of acquired epilepsy, and in nearly 30% of patients with seizures the cause is neurocysticercosis. An estimated 75 million individuals in Latin America alone are at risk of cysticercosis and taeniasis, of whom 400,000 are afflicted with symptomatic disease. Epidemiological studies using computed tomography (CT) in endemic settings have revealed asymptomatic brain calcifications in 10-20% of the general population.

Neurocysticercosis affects males and females equally and manifests predominantly in young adults, with a peak occurrence between 25 and 35 years of age; however, infection probably occurs earlier in life (5-15 years of age). The disease is endemic in most of the developing world, where all the condition favoring the transmission of this parasitosis, including warm climate, severe poverty, and illiteracy are combined. Together with the growing number of immigrants from endemic areas, there has been a recent increase in the number of patients with neurocysticercosis in the developed world.

3. Pathophysiologic Features

*T. solium* larvae are most common in the CNS, eyes, muscles, and subcutaneous tissue. *T. solium* may cause disease through any of three main mechanisms: (a) the presence of the parasite itself (ie, mass effect, obstruction); (b) the ensuing inflammatory response (ie, edema); and (c) residual scarring (ie, fibrosis, granulomas, and calcifications).

4. Clinical features

The clinical pleomorphism of neurocysticercosis is mainly related to individual differences in the number and location of the lesions within the CNS and to variations in the severity of disease activity. Seizures are the most common clinical manifestation of neurocysticercosis and may represent the primary or sole manifestation of the disease in almost 52 to 70% of patients. Seizures are more frequently observed in patients with parenchymal neurocysticercosis than in those with subarachnoid or ventricular disease. Recent data suggest that calcified cysticerci are not clinically inactive nor...
pathologically inert lesions, as they may cause recurrent seizures when parasitic antigens trapped in the calcium matrix are exposed to the host immune system due to a process of calcification remodeling (8).

Headache represents the second most common symptom (43%), followed by those caused by CSF obstruction (up to one-third of cases) (9). In contrast, signs of meningeal irritation are uncommon (<2% of cases) (1).

Focal neurological signs that vary according to the size, number and location of the parasites have been described in up to 20% patients with neurocysticercosis. These manifestations usually follow a subacute or chronic course resembling that of a brain tumor and are most often seen in patients with large subarachnoid cysts compressing the brain parenchyma (4). Stroke syndromes have also been described in about 3% of patients with neurocysticercosis. Some patients with neurocysticercosis develop intracranial hypertension associated or not with seizures or focal neurological signs. The most common cause of this syndrome is hydrocephalus, which may be either related to cysticercotic arachnoiditis, granular ependymitis, or ventricular cysts (4).

Some other patients with neurocysticercosis may present psychiatric manifestations ranging from poor performance on neuropsychological testing to a severe dementia (10). Spinal involvement, which accounts for only 1.5% of cases of neurocysticercosis (1). The intradural-extramedullary form is the most common, and clinical presentation often includes para or quadripareisis associated with sensory deficits (1).
**Fig. 1:** Life Cycle of Taenia Solium (Reprinted with permission of Garcia HH, Gonzalez AE et al. Neurocysticercosis some of the essentials. Practical Neurology 2006; 6: 288-297)

Fig. 2: Figure 2. Geographic distribution of Neurocysticercosis (Reprinted with permission of Garcia HH, Gonzalez AE et al. Neurocysticercosis some of the essentials. Practical Neurology 2006; 6: 288-297 with permission from BMJ Publishing Group Ltd)

© Reprinted with permission of Garcia HH, Gonzalez AE et al. Neurocysticercosis some of the essentials. Practical Neurology 2006; 6: 288-297 with permission from BMJ Publishing Group Ltd)
<table>
<thead>
<tr>
<th>Vesicular Stage (viable larva)</th>
<th>Colloidal Vesicular Stage (degenerating larva)</th>
</tr>
</thead>
<tbody>
<tr>
<td>- CT: smooth, thin-walled cyst, isodense to CSF, no edema, no enhancement</td>
<td></td>
</tr>
<tr>
<td>- MR: Cystic lesion isointense to CSF</td>
<td></td>
</tr>
<tr>
<td>May see eccentric scolex with hyperintense signal on DWI (13). The scolex also can be hyperintense on T1.</td>
<td></td>
</tr>
<tr>
<td>CT: hyperdense cyst fluid with surrounding edema</td>
<td></td>
</tr>
<tr>
<td>MR: cyst mildly hyperintense to CSF. Surrounding edema on FLAIR/T2. Thick cyst wall enhances. Enhancing marginal nodule (scolex). Also scolex with hyperintense signal on DWI in some cases.</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Granular Nodular (healing)</th>
<th>Nodular Calcified (healed)</th>
</tr>
</thead>
<tbody>
<tr>
<td>- CT: mild edema and involuting, enhancing nodule</td>
<td></td>
</tr>
<tr>
<td>- MR: Thickened, retracted cyst wall, edema decreases. May have nodular or ring enhancement</td>
<td></td>
</tr>
<tr>
<td>CT: Small, calcified nodule</td>
<td></td>
</tr>
<tr>
<td>MR: Shrunken, Ca++ lesion. T2 GRE useful to demonstrate calcified scolex. May show “multiple black dot” appearance</td>
<td></td>
</tr>
</tbody>
</table>

**Fig. 3:** CT and MRI findings of various stages of neurocysticercosis

© - Porto Alegre/BR
Findings and procedure details

We joined cases of neurocysticercosis from the radiology department of a tertiary hospital in the South of Brazil to illustrate the imaging findings of each phase of the cysticerci. We also joined a case from the pathology department and correlated with the magnetic resonance imaging findings. Neurocysticercosis has three most common forms: parenchymal, intraventricular and subarachnoid-cisternal. There is conflicting data in the literature about the most common of them. We considered parenchymal the most frequent according to our cases.

**Parenchymal Neurocysticercosis**

Neurocysticercosis most commonly manifests in the parenchyma of the brain and typically involves the cerebral hemispheres, with lesions commonly found at the gray matter-white matter junction, presumably resulting from deposition of the larvae in terminal small vessels of this region. Multiple lesions are usually present, although the number is highly variable, with solitary lesions as well as innumerable lesions in a miliary pattern occasionally encountered (11). Cerebellar, basal ganglia, or brainstem lesions may also be found in individuals with numerous lesions.

Neurocysticercosis has been classified into active and nonactive forms (1) on the basis of clinical presentation, results of CSF analysis (ie, hypoglycorrhachia, eosinophils in sediment, and cysticercus-specific immunoglobulin G antibody level), and imaging findings. The active forms include arachnoiditis with or without ventricular obstruction and vasculitis with or without infarction (1). On the basis of radiologic findings, neurocysticercosis is divided into five stages: noncystic, vesicular, colloidal vesicular, granular nodular, and calcified nodular (2, 14).

Noncystic neurocysticercosis (active) is asymptomatic with negative imaging findings (1). The earliest form of larval invasion is noncystic and is usually not detectable on imaging, because of frequent lack of edema at that stage (3).

Computed tomography (CT) and magnetic resonance (MR) imaging excel at detecting acute and chronic forms of neurocysticercosis and its complications. Four stages of neurocysticercosis lesions can be recognized on CT and MR imaging as they naturally evolve from the acute to the chronic form (Figure 3). Vesicular, colloidal vesicular, granular nodular and calcified nodular can be recognized in CT and MR images.
VESICULAR STAGE

In the first vesicular stage, the parasite cyst membrane is intact with little or no host response. In this phase, there is simple fluid within a cyst. The fluid has characteristics similar to cerebrospinal fluid on CT and all MR sequences (14). (Figure 4 and 5).

COLLOIDAL VESICULAR

As the cyst breaks down, fluid from the cyst becomes proteinaceous and leaks into the surrounding tissue, causing a strong immune response: the colloidal vesicular stage. On MRI, these lesions show a thick, well enhancing capsule following contrast with surrounding edema; on CT scanning they appear as hyperdense cysts (14). (Figures 6 and 7)

GRANULAR NODULAR

The cyst then further degenerates and oedema decreases in the granular nodular stage. MRI shows the transformation of these cysts into either nodular or mixed types. Nodular lesions have surrounding high signal on MRI, with a central nodule that has the same intensity as white matter. Mixed type lesions are high signal, with central hypointensity due to a mineralised scolex inside the cyst. On CT this gives the 'bull's eye sign' (14). (Figures 8 and 9).

CALCIFIED NODULAR STAGE

Finally, the cyst becomes calcified to form a granuloma in the nodular calcified stage. CT shows calcified lesions in the brain parenchyma. On MRI, these are micronodular lesions that are isointense on T1-weighted images and low signal on T2-weighted images (14) (Figure 10).

Intraventricular Neurocysticercosis
The intraventricular form of neurocysticercosis may be seen in 10% to 21% of neurocysticercosis cases. Although coexisting parenchymal lesions may be found, facilitating the radiologic diagnosis, up to 76% of patients may present with intraventricular lesion as the only detectable disease. This form of neurocysticercosis frequently manifests as a solitary intraventricular cyst (3). The fourth ventricle is the most common site (50%), followed by the lateral ventricles (35%), third ventricle (10%), and aqueduct (5%) (18). Isolated ventricular neurocysticercosis (ie, without subarachnoid disease) has been reported in one-third of cases (1). It often leads to obstructive hydrocephalus and ventriculitis due to ependymal inflammatory response (ie, granular ependymitis and subependymal gliosis) or adhesions due to prior ventricular infestation (3).

This form of neurocysticercosis may be challenging to identify on CT, especially if parenchymal calcified lesions are absent. The cysts are usually isointense to CSF on MR imaging with T2- and T1-weighted sequences, and only infrequently demonstrate hyperintensity to CSF on T1-weighted imaging and FLAIR sequences (3). (Figures 11 and 12) The wall of the cyst may occasionally be seen as a thin linear structure on T2-weighted imaging, or demonstrate contrast enhancement. Because many intraventricular neurocysticercosis cysts are isodense and isointense to CSF, they may be detected owing to presence of ventricular deformity, distention, and associated hydrocephalus (3). High-resolution MR cisternography sequences such as constructive interference in steady state (3D-CISS) may improve visualization of the intraventricular cyst and may resolve the internal scolex. Ependymal enhancement may be seen in cases with inflammatory involvement of the adjacent ventricle (3).

**Subarachnoid-cisternal neurocysticercosis**

The subarachnoid form of neurocysticercosis is the third most common manifestation of this disease after parenchymal and intraventricular forms, and has been found in 3.6% of neurocysticercosis patients. Neurocysticercosis of the subarachnoid spaces and the ventricular system is thought to result from hemotogenous dissemination of the larvae to these locations (3). Involvement of the basal cisterns is frequently identified and has been referred to as the meningobasal form (3). The suprasellar cistern, prepontine cistern, and ambient cisterns are among the most commonly involved. In these regions the scolex is often absent or degenerated, and multiple complex cysts may form, filling the involved cisterns with associated mass effect and distortion of adjacent structures including brainstem and cranial nerves. The multiloculated appearance of these fluid-filled lesions lacking internal scolex has been likened to a cluster of grapes, and is termed the "racemose" cysticercosis (3). However, as it is now recognized that degenerated
scolex can be found in these lesions, this designation has fallen out of favor (15) (Figure 13).

Cysticerci in this region are difficult to detect on CT. Even on MR imaging these lesions are often poorly visualized, owing to the isointensity of the lesions to CSF on all sequences similar to surrounding normal cisterns. Initially no enhancement may be detected within the basal cisterns, and the cysts may be seen primarily as a result of distortion of the involved cisterns and adjacent structures (3). The cyst walls may be identified as thin linear hypointensities on T2-weighted imaging. Identification and characterization of these lesions is greatly improved with high resolution MR cisternography sequences, such as 3D-CISS, through the basal cisterns. Cyst walls, degenerated scolex, and associated distortion of cranial nerves and vascular structures can be best visualized with this technique (3).
Fig. 5: Vesicular Stage. A. Axial T2 MR shows a left parietal cystic lesion (thick arrow) with a scolex (thin arrow). B. Axial FLAIR MR. Note that the cyst (thick arrow) is isointense to cerebrospinal fluid (CSF).

© - Porto Alegre/BR
Fig. 4: Vesicular Stage. A. Axial T2 MR shows a cyst in the right temporal lobe (thick arrow) in the same patient. The round black structure within the cyst (thin arrow) represents the scolex. B. Coronal T2 MR demonstrates a cyst in the right temporal lobe in another patient (thick arrow).

© - Porto Alegre/BR

Fig. 6: Colloidal vesicular stage. A. Axial T2 MR demonstrates a left parietal lesion with edema. B. Axial gadolinium-enhanced T1 MR shows wall enhancement.

© - Porto Alegre/BR
Fig. 7: Colloidal vesicular stage. A and B. Axial T2 and FLAIR MR shows a parenchymal hyperintense parenchymal cyst in the right middle frontal gyrus. C. Axial DWI MR shows the scolex with hyperintense signal on diffusion. D. Axial gadolinium-enhanced T1 MR shows wall enhancement.

© - Porto Alegre/BR
Fig. 8: A. Coronal T2 MR shows a left parietal lesion with central hypointensity and peripheral edema (arrow). B. Axial gadolinium-enhanced T1WI MR in the same patient shows thick cyst wall enhancement (arrow).

© - Porto Alegre/BR

Fig. 9: Granular Nodular Stage. A. Axial FLAIR MR image shows left superior temporal gyrus lesion (arrow) with thickened and retracted wall. Surrounding edema regressed partially. It represents progressive involution of the cyst . B. Axial gadolinium-enhanced T1WI MR in the same patient shows thin cyst wall enhancement (arrow).

© - Porto Alegre/BR
Fig. 10: Calcified Nodular Stage. A and B. CT demonstrates multiple calcified nodules in two different patients. C and D. Axial T2 and T2 GRE MR shows multifocal "black dots" in the sulci, parenchyma and left frontal horn related to the nodular calcified stage. It is important to remember that different lesions may be at different stages in the same patient.

© - Porto Alegre/BR
Fig. 11: Intraventricular Neurocysticercosis of the fourth ventricle: A and B. Sagittal T2 and T1 MR image demonstrates a large complex cystic lesion within the fourth ventricle with thin linear walls (arrows in A), small enhancing nodule (arrow in B), and internal cyst fluid predominantly isointense to CSF. C and D. Coronal T2 and Axial FLAIR MR shows supratentorial hydrocephalus and edema around the lesion.

© - Porto Alegre/BR

Fig. 12: Intraventricular neurocysticercosis: Axial T2 MR shows a cystic lesion surrounded by mild edema in the anterior horn of the left lateral ventricle (red arrows). There are surgical alterations close to the posterior horn of the right lateral ventricle to treat the hydrocephalus (blue arrow).

© - Porto Alegre/BR
**Fig. 13:** Subarachnoid-cisternal neurocysticercosis: A and B. Axial T2 weighted image demonstrates multiple cystic lesions filling suprasellar cistern (red arrows) and ambiens/quadrigeminal cisterns (yellow arrow) without any visible scolex. Lobulated cysts with no mural nodule and signal intensity similar to that of CSF was found in the cerebello-pontine angle (blue arrows) and pontine cistern (green arrows). CSF serology (anti-TaeniaSoliumantibody) was positive. C and D. Autopsy specimen showing gross morphology of neurocysticercosis lesions.

© - Porto Alegre/BR
Conclusion

6. Conclusion

There are several ways to classify the cysticerci in the CNS, and it will depend on many factors. These patterns will change the management and prognosis of the patient. Therefore, it is important for the neuroradiologist recognizes them.

The radiologist should always remember that complex parasitic cysts may mimic brain tumor and that the clinical presentation can help in the differential diagnosis. Cysticercosis is endemic in many countries, especially in Latin America, parts of Asia, India and Africa, and can occur in any age. It is the most important parasitic disease of the human central nervous system and constitutes a public health challenge for most of the developing world. The treatment will change according to the disease stage and type.
Personal information

L. G. Campos¹,², J. Zampieri¹, F. D. S. Souza¹, M. G. Longo¹, J. Duarte¹, J. A. Perez¹, L. Vedolin¹; Porto Alegre/BR

1. Department of Neuroradiology of Hospital de Clínicas de Porto Alegre

2. cgillian@yahoo.com
References


