Neurocysticercosis (NCC)

Neurocysticercosis is the most common parasitic infection of the CNS.

Key concepts

● intracranial encystment of larva of Taenia solium (pork tapeworm).

● the most common parasitic infection of the CNS.

● neurological symptoms: seizures or progressive intracranial hypertension.

● occurs from ingesting the parasite's eggs, not from eating infested meat.

● characteristic imaging finding: low density cysts with eccentric punctate high density (the scolex= tapeworm head). Hydrocephalus is common.

● medical treatment: all patients get steroids. Start antihelmintic drugs (praziquantel or albendazole) when no signs of intracranial hypertension.

● biopsy sometimes needed for diagnosis. Surgery: may be required for spinal, intraventricular or subarachnoid cysts (more refractory to medical therapy) or for giant cysts (>50 mm) when intracranial hypertension persists despite steroids

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Extraneural cysticercosis causes no major symptoms but neurocysticercosis and ophthalmic cysticercosis are associated substantial morbidity.

The parasites spread through the bloodstream and it may locate almost anywhere in the body. In CNS, cysticercosis frequently involves the cerebral hemispheres, ventricles and basal cisterns, subarachnoid space and spine whereas the most frequent location is in the cerebral hemispheres, mainly at the junction of gray and white matter [1) 2)].

Moreover, racemose cysticercocis is common in the cisternal or subarachnoid space [3)].

Epidemiology

Neurocysticercosis is common in less developed countries but is increasingly reported in more developed countries because of immigration of people from endemic areas [4)].

NCC should be considered as a possible diagnosis for patients suffering from hydrocephalus when they originate from or have traveled in endemic areas, MRI of the spine is mandatory to search for intraspinal lesions [5)].

Classification

Neurocysticercosis (NCC) will be developed when onchospheres reach the central nervous system which can be classified in parenchymal and extraparenchymal, depending on the location of the cysts.

Cerebral hemispheres are the most commonly affected, mainly at the gray-white matter junction and cysts can be also found in the ventricles, basal cisterns and subarachnoid spaces.

In the posterior fossa, neurocysticercosis usually involve the fourth ventricle, cerebellopontine angle cistern, cisterna magna and rarely, the cerebellum [6) 7)].

see intraventricular neurocysticercosis

see cerebellar cysticercosis

cerebellopontine angle cysticercosis....

Racemose neurocysticercosis

Clinical Features

Neurocysticercosis causes symptoms in most patients years after CNS infection by the parasite. Clinical manifestations result from inflammatory response to cyst degeneration, mass effect, obstruction of CSF pathway or residual scarrin [8) 9)].

Clinical manifestations are nonspecific and varied according to locations, number and size of cysts, and host immune response to the cysticerci [10)].

Epileptic seizures are the most frequent presentation of parenchymal neurocysticercosis but large cyst may produce mass effect. Extraparenchymal neurocysticercosis in the ventricles and basal cisterns can be present with hydrocephalus or intracranial hypertension by mechanical obstruction of CSF pathway, either by the cysts or by an inflammation. A large multicystic cerebellar mass cab cause hydrocephalus and intracranial hypertension by mechanical obstruction of fourth ventricle [11)].

Diagnosis

In 2017, diagnostic criteria for NCC were edited and organized in three categories with many levels of certainty: absolute, neuroimaging and clinical/exposure criteria. Histological confirmation of parasites, evidence of subretinal cysts, and demonstration of the scolex within a cyst are absolute criteria for NCC diagnosis. Cystic lesions without scolex, multilobulated cysts and calcifications are categorized as neuroimaging major criteria [12) 13)].

Imaging findings vary with development stage of neurocysticercosis or host response and lesions may be at different stages in same patient [14) 15) 16)].

On imaging studies, viable cysts appear isodense with the cerebrospinal fluid and there are no contrast enhancement with little or no evidence of perilesional inflammation (vesicular stage). As the

parasites degenerate, the cysts show pericystic contrast enhancement with perilesional edema (colloid stage). In later stage, they have nodular or ring enhancement after the administration of contrast, like in our case (granular-nodular stage). Finally, the cysts are not detectable or become calcified lesions (calcific stage).

Differential diagnosis

A variant of pilocytic astrocytomas and his recognition may prevent an unnecessary workup to exclude other etiologies such as parasitic infection (ie, cysticercosis) or cystic metastatic disease [17].

Atypical brain metastases [18]

Include abscess, tuberculosis, metastases and other parasitic diseases [19].

However, differential diagnosis between cysticercosis and other diseases may be difficult because clinical manifestations are not specific and some neuroimaging findings are not pathognomic. Therefore, immunological tests and epidermiological data as well as neuroimaging studies are helpful to diagnose neurocysticercosis [20].

Treatment

Neurocysticercosis Treatment.

Case series

Clinical, demographic and neuropathological findings of histologically confirmed cases of Neurocysticercosis (NCC) causing drug resistant epilepsy between 2005-2019 were reviewed. NeuN, GFAP, phosphorylated neurofilament, vimentin, CD34 for glial/ neuronal alterations, and Masson trichrome, Luxol Fast blue for evidence of fibrosis/ demyelination was used to determine cause of epileptogenesis.

There were 12 cases of NCC associated with dual/ double pathology, which constituted 3.02 % (12/398) of all the operated DRE. [Age range: 17-37y, Male:Female = 1.4:1]. Seizure duration ranged from 3-32y, with seizure onset between 4-27y. On MRI, lesions were of variable signal intensity on T1 and isointense on T2 with blooming on GRE/ SWI, and CT revealed calcification. Majority (11/12) had associated hippocampal sclerosis (HS) type 1 (dual pathology), localised to the same side as cysticercal cyst, suggesting it may be involved in the pathogenesis of HS. Ten had single cysticercal lesion involving ipsilateral hippocampus in 6, parahippocampal gyrus in 2, amygdala and temporal lobe in 1 case each. One had multiple NCC located in bilateral frontal, parietal and ipsilateral hippocampus. Adjacent cortex around the NCC evaluated in 6 cases, revealed inflammation, gliosis, axonal disruption/ beading, and variable synaptic/ neuronal dystrophic changes. There was a single case of NCC with Focal cortical dysplasia (FCD) type IIb (double pathology). In 11/12 cases Engel's post-surgery outcome was available with all having class I outcome.

Hippocampal sclerosis (HS) was most common pathology associated with cysticercosis (Dual pathology), localised ipsilateral to the cysticercal cyst, suggesting that HS is a secondary/

epiphenomenon. Perilesional changes such as inflammation, gliosis, dystrophic synaptic and axonal pathology play a role in inducing or perpetuating the epileptiform activity. The association of FCD IIb with NCC in one case is likely to be a chance occurrence [21)].

Twenty-six patients with cysticercosis of the brain parenchyma were treated with the antihelmintic agent praziquantel (50 mg per kilogram of body weight daily for 15 days). During treatment a strong inflammatory reaction occurred, as evidenced by increased protein and cells in the cerebrospinal fluid. This finding correlated with headache, exacerbation of neurologic symptoms, and edema and inflammation around cystic lesions. After three months of treatment all patients had improved clinically, and 13 (50 per cent) were asymptomatic. The total number of cysts on CT scans had decreased from 152 at the beginning of treatment to 51, and the mean diameter of cysts was reduced by 72 per cent. CT scans showed improvement in 25 of the 26 patients, with total remission of all cysts in nine. Seventeen control patients followed with CT studies for a mean of 9 +/- 2 months had no spontaneous remission of lesions, and in many cases the scans showed worsening during the observation period. The results indicate that praziquantel is effective in cysticercosis of the brain parenchyma [22)].

Case reports

Nieto Moragas et al., from the Hospital Universitari Germans Trias i Pujol, Universitat Autònoma de Barcelona, Badalona, Spain. reported a 40-year-old woman from Ecuador who presented to the emergency department (ED) with headache, vomiting, drowsiness, confusion and incoherent speech. She lives in Spain since 2001 and she carries a ventriculoperitoneal shunting since 1993, placed in Ecuador because an obstructive hydrocephalus of unknown origin. In 2013, she consulted repeatedly the ED because hydrocephalus valve mismatches requiring a valve replacement. Laboratory test in peripheral blood showed C-reactive protein level of 34.90 mg/dl and 7800 neutrophils/dl. Cerebro-spinal fluid (CSF) analysis showed 260 red blood cells/l, 300 leukocytes/l with a differential of 42% of eosinophils, a protein level of 1.5 g/l and positive xanthochromia. Gram stain of CSF showed no microorganisms and was inoculated onto chocolate blood agar and blood agar plates and incubated in an aerobic atmosphere at 37 ◦C with negative result after 48 h. Computed tomography (CT) confirms valve dysfunction excluding left ventricular and showed some parenchymal and periventricular calcifications. The patient underwent emergency valve replacement with proximal catheter placement. MRI postintervention showed some punctate supratentorial calcifications and two spherical multi-septate lesions with a CSF similar density placed periventricular in the left ventricle. One ofthe multi-septate lesions supposedly responsible of the symptoms was removed surgically and some histological cuts were made of it.

A sample of CSF was sent to the Parasitology Unit of the Instituto de Salud Carlos III (Madrid, Spain) to detect by enzyme-linked immunoassay Taenia solium antigen and immunoglobulin G (IgG). Both antigen and IgG against T. solium tests were positive and it was confirmed with a polymerase chain reaction (PCR) targeted to a T. solium DNA on CSF.Parenchymal and periventricular calcifications, two spherical multi-septate lesions, the presence of anticysticercal antibodies in CSF and been residing in an endemic country, where the patient had already showed intracranial hypertension, lead to the neurocysticercosis diagnosis. Antiparasitic drugs were no recommended at this stage of the disease and the treatment with parenteral dexamethasone was needed till a progressive neurological improvement. Sequelae included language disorder and slow mental activity that improved with neurocognitive rehabilitation [23)].

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