CLINICAL VIGNETTE

Recurrent Racemose Neurocysticercosis

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Introduction

Neurocysticercosis (NCC) is the most common subacute and invasive parasitic infection of the central nervous system caused by the encysted larval stage of the pork tapeworm Taenia Solium.[1,2] The larvae, in form of cysts, can infect any organ, but is most commonly found in the CNS, the eye, and the muscle.[2] If the infection is found within the grey-white matter junction of the cerebral hemispheres, it is considered intra-parenchymal subtype, while an extra-parenchymal subtype, also known as racemose, refers to infections involving the subarachnoid, meningeal, and intraventricular space.[3] Racemose NCC is characterized by the presence of space occupying larval cysts within the subarachnoid basal cisterns or sylvian fissures and is often described as a multicystic grape-like cluster on imaging,[4] which is a key diagnostic feature. As the more severe subtype, racemose NCC is associated with a characteristic absence of the scolex in the fluid filled bladder of the cyst,[2,5] lack of contrast-enhancement or edema on imaging, and higher mortality (30-80%) with decreased response to treatment.[2] While NCC can be observed worldwide, it is primarily endemic to underdeveloped regions in Latin America, Africa, Asia, and Eastern Europe and often associated with poor sanitary conditions.[1,4]

The clinical presentation of NCC can vary, depending on the size and location of the parasitic load and the host immune system. Early symptoms include headaches of varying degree, psychiatric changes, rapidly progressing dementia, followed by intracranial hypertension, vasculitis, seizures, stroke, and life-threatening hydrocephalus.[1] Since there is significant heterogeneity in presentation,[1] it is estimated that NCC is widely underdiagnosed and some studies suggest that up to 30% of epilepsy in endemic regions may be due to untreated cases of NCC.[3]

Early recognition is key to reducing mortality and morbidity. The treatment of choice is extended therapy with Albendazole for 4 weeks, due to its improved CSF penetration as compared to Praziquantel.[6] Some cases, especially racemose NCC, require coadministration of anti-inflammatory agents such as corticosteroids as well as intraventricular shunt placement due to the hydrocephalic obstruction created by the larval cysts in the subarachnoid space.[2,5]

Case Presentation

A 52-year old male with known history of racemose Neurocysticercosis (rNCC) treated medically as well as surgically with ventriculoperitoneal (VP) shunt placement, presented with rapidly worsening forgetfulness, myalgias, headaches, nausea, weight loss, blurred vision and right sided hearing loss over 2-3 months. He was followed closely as an outpatient by Infectious disease and Neurology. On the day of admission, he presented to ID clinic, with stable vital signs, and without signs of meningeal irritation. However, his physical exam was notable for new neurological focal deficits of decreased right hearing loss to finger rub and an unsteady gait during tandem walk. CT head without contrast completed 1 week prior to the clinic visit, showed intact VP shunt, stable focal calcifications consistent with known history of rNCC but an increase in size of the fluid density involving the right cerebellopontine angle cistern in comparison to the prior year's CT scan. Due to the new onset neurological defects and CT head changes suggesting active rNCC, he was admitted for IV antiparasitic treatment with concurrent steroid administration for prevention of intracranial hypertension secondary to cyst degeneration and host inflammatory response.[7,8] Admission, labs included WBC of 18.6, platelet count of 468, sodium of 131, BUN of 26 and blood glucose in the 300s. Brain MRI confirmed the presence of active rNCC with larval cysts of varying developmental stages (Figure 1).

Her diagnosis of recurrent racemose Neurocysticercosis and encephalitis caused by Taenia Solium confirmed and Neurosurgery determined the patient was not a candidate for surgical intervention. He received antiparasitic treatment with IV albendazole 30mg/kg/day and PO Dexamethasone 16mg daily. His neurologic symptoms improved after day 5 and repeat MRI of discharge revealed decreased size of the cystic lesion at the right cerebellopontine angle reflecting treatment response. He was discharged on Calcium-Vitamin D twice a day, Omeprazole 20mg daily, trimethoprim sulfamethoxazole MWF, and a Dexamethasone taper with close clinic follow up with Ophthalmology, Neurology, Neurosurgery and Infectious disease.

One month after discharge, the patient presented with headaches, blurry vision, fatigue, dizziness and new onset right ptosis and eyelid edema. His hearing was improving. Ophthalmology felt the blurry vision was likely refractory after negative funduscopic exams and serial CT scans showing lesion stability. Potential surgical removal of isolated cysts not responding to medical treatment, will closely monitored by Neurosurgery.

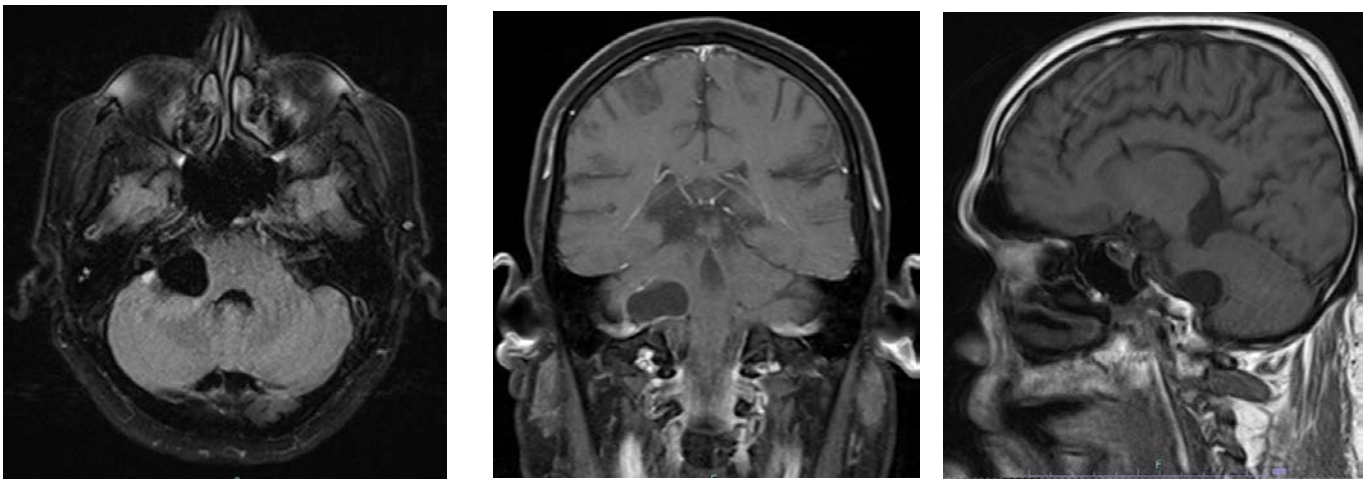

Figure 1. Axial, coronal, sagittal T1-weighted magnetic resonance imaging demonstrating a lesion involving the right cerebellopontine angle cistern extending into the right premedullary cistern with thin rim enhancement and surrounding FLAIR hyperintensity in the right cerebellum with mass effect on the right 7th and 8th cranial nerve and right anterior medulla.

Discussion

This patient with recurrent racemose NCC, likely due to insufficient eradication of his original infection many years ago. There is no current consensus regarding optimal treatment of rNCC, including optimal antiparasitic, duration, dosage, or combination therapies with surgical intervention.[9] Existing literature suggests less aggressive antiparasitic treatment and subsequent shunt infection directly lead to poor outcomes.[10] However, shunt placement alone is not a predictor of treatment failure, as >80% of racemose NCC cases undergo a shunt placement during their treatment.[11] Additional surgical intervention is usually reserved for severe cases after medical treatment fails or the patient experiences significant neurological decline. High morbidity associated with surgical complications, include arachnoiditis and adherence of evacuated cyst walls to cranial nerves and arteries.[12]

Chart review revealed the patient had complained of headaches for 6 years before the current presentation and 2 years after shunt replacement, Prior CT imaging revealed new cystic growth but unfortunately the patient was lost to follow up. He had also been seen by Cardiology for left sided chest pain four years ago which then developed into global body aches, which was possibly due to ongoing inflammation causing vasculitis and cerebrovascular complications often seen in NCC.[11] He underwent a negative exercise stress test but did not have Angiography or MRA imaging, which may have revealed segmental narrowing of large vessels within the basal cisterns and sylvian fissures as commonly seen in rNCC,[11,13,14] which can lead to cranial nerve dysfunction due to fibrous entrapment.[15,16] These early symptoms – headache, chest pain, body aches – had been suspicious for recurrent rNCC infection and repeat head imaging had shown gradual increase in size of known cystic lesions. Medical treatment could have been initiated earlier, if the patient had not been lost to follow up.

Conclusion

Racemose Neurocysticercosis is a rare version of a Taenia Solium infection affecting the CNS spaces, where it commonly presents in the subarachnoid basal cisterns and sylvian fissures as space occupying larval cysts of varying developmental stages. Due to the high mortality and morbidity, early treatment is of the utmost importance, which requires vigilence by providers. Early symptoms of rNCC can be nonspecific – headaches, body aches, vascular and neurologic issues – and therefore rNCC can mimic many other more common conditions such as stroke, dementia, and other neurological disorders. In addition to early detection via head imaging and/or ELISA antibody tests for Taenia Solium, close follow up after treatment is important in ensuring eradication of the infection and preventing recurrence years later. This case serves as an example of how a complex disease process, presenting in a vulnerable patient population with limited access to healthcare, can still have improved outcomes with a multidisciplinary approach and close outpatient care.

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