G55 Headache and Sudden Death in a Young Adult: An Unexpected Finding at Autopsy

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After attending this presentation, attendees will have gained a basic understanding of the different ways humans can contract Taenia solium, the pork tapeworm, and the different diseases that can develop from infection with this parasite, emphasizing involvement of the central nervous system.

A 33-year-old white female presented with a history of severe headaches, nausea, vomiting, and dizziness. The headaches, which were unassociated with fever, began in February 2006. A work up for meningitis including examination of cerebral spinal fluid was negative, resulting in a diagnosis of migraine headaches. The headaches and accompanying symptoms, however, persisted. Following an episode of one of the headaches, she was found unresponsive in bed and was unable to be resuscitated.

Autopsy examination revealed the presence of cerebral edema with tonsillar herniation. The posterior horns of the lateral ventricles in the temporal region were dilated. The fourth ventricle was dilated and obstructed by a colloidal appearing cystic structure that measured 1 cm in maximum dimension. The remainder of the autopsy examination was unremarkable. Blood cultures were negative, vitreous electrolytes showed a normal postmortem pattern and were unremarkable, and toxicology testing was negative.

Histologically, the brain showed an intense mononuclear infiltrate composed of lymphocytes and plasma cells that surrounded a homogenous amorphous eosinophilic structure in the ventricle. The choroid plexus focally showed a similar intense inflammatory reaction. Ependymitis and mononuclear perivascular cuffing were also present. The cystic structure seen grossly in the fourth ventricle had a wall that was composed of three layers: an eosinophilic outer cuticle with the grapelike appearance, a single layer of subcuticular cells, and a myxoid cytoplasm containing tubular structures. These findings are consistent with intraventricular neurocysticercosis.

Upon further questioning of the family, it was learned that the decedent had immigrated to the United States from Mexico 12 years ago. Her family in Mexico owned a restaurant type business where pigs were raised and slaughtered on the property. Prior to her recent complaints of headache, the decedent had been in good health.

Cysticercosis is the most common parasitic disease affecting the central nervous system. It is endemic in Latin America, India, China, Southeast Asia, and sub-Saharan Africa. It is estimated that up to 90 percent of patients with cysticercosis have central nervous system involvement. Sole involvement of the central nervous system is termed neurocysticercosis (NCC). The disease is reported in all age groups, but most cases present in the third and fourth decades. Reliable data regarding the incubation period are lacking, but it is estimated that months to decades can pass between initial infection/exposure and the subsequent development of neurological symptoms.

The pork tapeworm Taenia solium causes the infection. Humans can be either the definitive host or the intermediate host and thus two different diseases are recognized. Infection with the larval form as would occur from eating contaminated undercooked meat leads to taeniasis. The larva hatches in the small intestine and develops into tapeworms. In this situation humans are the definitive host because they harbor the adult form of the parasite – the tapeworm. In contrast, when humans harbor the larval form of the parasite, they are the intermediate host and the disease is called cysticercosis. This occurs when *T. solium* eggs are ingested. Sources of eggs include contaminated food and water, fruit, or vegetables fertilized with contaminated human or pig feces and contact with individuals harboring the tapeworm and shedding the eggs. The disease may be contracted from fomites because the eggs are resistant to environmental conditions.

In the small intestine the eggs hatch releasing oncospheres. The oncosphere penetrates the intestinal mucosa, travels to the pulmonary circulation, and is disseminated systemically. The oncosphere, which develops into the larval form, may reach several different organ systems, but seem to have a predilection for the central nervous system, skeletal muscle, subcutaneous tissue, and eyes. Central nervous system involvement most commonly involves the parenchyma and may cause seizures.

Involvement in the central nervous system may be extraparenchymal affecting the ventricles, subarachnoid space, eyes, and spinal cord. It is estimated that between ten to 30 percent of patients with NCC have intraventricular cysts. Ventricular cysts can be attached to the ependyma or float freely migrating throughout the cerebral spinal fluid pathways**.** Unlike parenchymal cysts, which are typically multiple, ventricular cysts tend to be solitary and ventricular involvement typically occurs without accompanying parenchymal cysts. Ventricular cysts are more likely to be symptomatic than parenchymal

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cysts. Ventricular cysts can cause hydrocephalus either by blocking the flow of cerebral spinal fluid or by producing ependymitis with scarring, obstruction, and ventriculitis. Individuals having involvement of the ventricle most commonly present with signs of increased intracranial pressure such as headache, nausea, and vomiting. These symptoms are commonly attributed to migraine or tension headaches.

NCC is generally a chronic disease whose natural progression includes four stages: vesicular, colloidal, granular/nodular and calcified. Symptoms typically develop as the parasite begins to die losing its ability to control host defenses. The ensuing inflammatory response results in degeneration of the larva and the formation of a granuloma.

Making the diagnosis in a clinical setting can be difficult because the clinical manifestations are variable and nonspecific and depend on the number and location of cysts and the host's immune response. Proposed diagnostic criteria incorporate absolute, major, minor, and epidemiological criteria. Interpretation of the criteria allows for two degrees of diagnostic certainty – definitive and probable.

No reliable information is available regarding mortality rates. Large autopsy series from endemic areas suggests that the majority of cases are asymptomatic making calculation of the true incidence and prevalence difficult. It is estimated that 50 million people are infected worldwide. The disease carries a high cost in morbidity. In endemic countries NCC may be responsible for 50 percent of adult-onset seizure disorders and those with intraventricular and subarachnoid involvement can develop complications such as vasculitis and hydrocephalus. The annual treatment cost in endemic areas is estimated to be close to $90 million per year. In the United States, there are more cases of imported NCC than in all other developed countries combined, and the annual treatment cost is estimated at $9 million per year. The disease is generally encountered in the southwest United States and among Hispanic immigrants. The increasing number of reported cases in the United States is most likely due to increased immigration and travel to endemic areas. The long latent period and variable clinical presentations make it go undetected or unrecognized clinically. Although autopsy studies suggest that NCC is most commonly an incidental finding, the disease can result in death. NCC should be considered in the differential diagnosis of calcified, nodular, or cystic lesions of the central nervous system.

Neurocysticercosis, Headache, Central Nervous System

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