G15 Case Report – Sudden Death Due to Cystic Tumor of the Atrioventricular Node

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The goal of this presentation is to show the importance of including cystic tumors of the atrioventricular node as one of the main differential diagnoses in cases of sudden death, especially in the context of congenital heart block with the adjunct of performing a thorough examination of the cardiac conduction system.

This presentation will impact the forensic community and/or humanity by demonstrating how this type of tumor will be missed if routine sections of the AV node are not submitted in cases of sudden death, especially those involving young, healthy individuals, and it should always be considered in cases of sudden death in the context of congenital heart block.

Cystic tumor of the atrioventricular (AV) node is a benign, congenital, cystic mass located at the base of the atria septum in the region of the AV node. Although cystic tumor of the AV node is the most common intracranial tumor causing sudden death it is considered a rare neoplasm with less than 100 cases reported in the literature to date.

A case of a woman in her 30's who was diagnosed as an infant with complete heart block is reported. A permanent pacemaker with epicardial leads was subsequently placed. She functioned normally with the exception of exercise related shortness of breath. She underwent several pacemaker changes throughout her life. She had a very active lifestyle. She was in her usual state of good health when she experienced a sudden witnessed collapse at her workplace. Her initial cardiac rhythm at the scene was pulseless electric activity with appropriate pacemaker discharge. She was pronounced dead at the hospital after unsuccessful resuscitative measures.

At autopsy, examination of the cardiovascular system disclosed a 550gram heart. The coronary arteries had a normal distribution and were free of atherosclerosis. The pacemaker leads were appropriately positioned. No gross lesions were visible on examination of the cardiac conduction system.

Microscopic examination of the myocardium showed hypertrophic myocytes, focal interstitial fibrosis, and focal contraction band necrosis. Sections from the region of the AV node showed a proliferation of cells forming nests, cysts, and glands of variable size and shape measuring a minimum of one centimeter. The cell population ranged from those resembling transitional cells and those with squamous differentiation to cuboidal cells and clear, sebaceous-appearing cells.

Cystic Tumor of the Atrioventricular (AV) node has been called one of the "smallest tumors causing sudden death." When symptomatic the majority of patients present with complete heart block. The diagnosis of heart block in patients with AV nodal tumors may be made at birth or as late as the ninth decade of life. It has a female predominance. The majority of known cases are diagnosed at autopsy although a few reported cases have been diagnosed during life and treated. Pacemaker is the first line of therapy; however they are not always effective, as seen in this case. Although rare and histologically benign, cystic tumors of the AV node are the most common intracardiac neoplasms causing sudden death. They are located in the AV nodal region because this is an area of embryologic fusion and therefore prone to accidental incorporation of embryologic structures. The mechanism of death is related to its intracardiac location, which can precipitate conductive and hemodynamic abnormalities. Cystic tumors of the AV node are rarely seen grossly, but when visible it is seen as an elevated nodule above the septal leaflet of the tricuspid valve. Most of the time they are first identified microscopically. This lesion is characterized by multiple microcysts, gland like structures, and nests of epithelioid (occasionally squamoid) cells within a fibrous stroma. Previously thought of as a mesothelioma of the AV node, this lesion has since been shown convincingly to represent an endodermal heterotopia.

The tumor will be missed if routine sections of the AV node are not submitted in cases of sudden death, especially those involving young, healthy individuals. It should always be considered in cases of sudden death in the context of congenital heart block and congenital fibrosis of the AV node.

Cystic Tumor of the AV Node, Sudden Death, Heart Block

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