Letters to the Editor

Cystic Tumor of the Atrioventricular Node: A Rare Cardiac Pseudoneoplastic Lesion

To the Editor.—We read the outstanding article by Miller and Tazelaar1 recently published in the Archives of Pathology & Laboratory Medicine about the main features of 5 cardiovascular pseudoneoplasms (inflammatory myofibroblastic tumor, hamartoma of mature cardiac myocytes, mesothelial monocytic cardiac excrescences, calcified amorphous tumor, and lipomatous hypertrophy of the atrial septum). Although lacking malignant potential, these lesions can be associated with morbidity and occasional mortality, and their recognition is important for appropriate patient management.

We describe the main features of the cystic tumor of the atrioventricular node (CTAVN), another pseudoneoplastic lesion that may cause sudden death. First described in 1911,2 its controversial histogenesis resulted in several names3,4 such as mesothelioma, lymphangioendothelioma, endodermal inclusion, hamartoma, and so forth. We showed that CTAVN and solid cell nests of the thyroid gland are identical structures and indicated that CTAVN is an ultimobranchial heterotopia secondary to an alteration in cardiac neural crest cell development.3 Its association with several congenital abnormalities3,4 and possible familial occurrence5 suggest a genetic defect in cardiac neural crest cell development.3

Grossly, the heart can appear normal or show small cysts in the triangle of Koch.6 Cystic tumor of the atrioventricular node runs from the ostium of the coronary sinus in the right atrium to the membranous septum, along the top of the tricuspid valve septal leaflet. Histologically, it is composed of squamous solid cell nests and cystic structures with a minor proportion of neuroendocrine cells3–10 (Figure, A through E). Squamous cells (main cells) are polygonal to elongated with centrally located oval nuclei and eosinophilic cytoplasm. The neuroendocrine cells show clear cytoplasm and compact nuclei. Cysts usually contain mucous substances and are lined by mucinous, ciliated, transitional, and/or squamous cells. Sebaceous and goblet cells are described. Mitotic figures are usually absent. The main cells are immunoreactive for cytokeratins (cytokeratin 7 and clones CAM 5.2, AE1/AE3, and 34βE12), epithelial membrane antigen, carcinoembryonic antigen, carbohydrate antigen 19.9, p63, bcl-2, and galectin-3 with no reaction for vimentin, cytokeratin 20, CD31, factor VIII–related antigen, thrombomodulin, Wilms tumor 1, or calretinin. The neuroendocrine cells...
are positive for cytokeratin 7, CAM 5.2, and AE1/AE3, epithelial membrane antigen, carinoembryonic antigen, thyroid transcription factor 1, calcitonin, serotonin, chromogranin, and synaptophysin. Ultrastructurally, the main cells are characterized by cytoplasmic tonofilaments and desmosome-like structures, while the neuroendocrine cells showed electron-dense secretory granules. An angiomatous variant of CTAVN has also been reported.11

One patient was reported to have developed symptoms during her pregnancy and the tumors cells were positive for estrogen and progesterone receptors suggesting that the CTAVN might have been progressive during the pregnancy stage. Another case treated with long-acting gonadotropin-releasing hormone decreased in size; however, with a combination of estrogen and progesterin, the tumor increased in size and progressed even after the discontinuation of all therapy. These findings suggest that there might be a correlation between the progression of CTAVN and the levels of circulating hormones.8 However, we were unable to find positivity for estrogen and progesterone receptors in this tumor type.5 Clinically, CTAVN can cause heart blockage and sudden death as a result of ventricular tachycardia or ventricular fibrillation.5,6,9 The mean age of presentation is 38 years (range, birth to 78 years), with a female to male ratio of 3:1.9 Most of these tumors were diagnosed postmortem3,5; some of them as an incidental finding, because they were clinically silent. Antemortem diagnosis and successful excision of CTAVN was reported in 7 cases,6,8 and in 1 of them the tumor was an unexpected finding in an explanted heart.10 Cystic tumor of the atrioventricular node is an important differential diagnosis in young patients with syncopal attacks and varying degrees of heart blockage.10 It is also important to note that in patients with CTAVN, pacemaker implantation does not prevent death because these patients are at risk of lethal arrhythmias unconnected to the heart blockage.6 Complete surgical resection is essential, even if a subsequent pacemaker implant is required.6

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This study was supported by grant PI060290 from Instituto de Salud Carlos III (Ministry of Health and Consumer Affairs), Spain (Dr. Cameselle-Teijeiro). The authors have no relevant financial interest in the products or companies described in this article.

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Prepare Now for the CAP ’11 Abstract Program

Plan now to submit abstracts and case studies for the College of American Pathologists (CAP) 2011 meeting, which will be held September 11th through the 14th in Grapevine, Texas. Submissions for the CAP ’11 Abstract Program will be accepted from:

Monday, January 31, 2011 through Friday, April 1, 2011.

Accepted submissions will appear in the September 2011 issue of the Archives of Pathology & Laboratory Medicine. Visit the ARCHIVES Web site at www.archivesofpathology.org and also the CAP ’11 Web site at www.cap.org/cap11 for additional abstract program information as it becomes available.