G36 Coronary Artery Anomalies and Sudden Death: Two Case Reports in Young People

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The goal of this presentation is to present to the forensic community two cases of sudden death due to coronary artery anomalies by autoptic and clinicopathologic findings and histological studies.

This presentation will impact the forensic community and/or humanity by presenting to the forensic community two cases of sudden cardiac death due to rare coronary artery anomalies in young people. These two cases have been studied by means of autoptic and histological examinations. It would be an important contribute to the scientific community for the diagnosis of one of the possible causes of sudden death in infant and young people.

Coronary artery anomalies may cause sudden death. This presentation discusses the clinicopathologic features, the autoptic findings and microscopic features in two cases with different coronary anomalies.

Case 1: A 13-month-old infant, white male, was found unresponsive in his crib. The infant was transported to the hospital, where resuscitative efforts were unsuccessful, and the infant was pronounced dead. Previous clinical history: at the age of 6 months numerous episodes of apnea's crisis, cyanosis and convulsion and at the age of 7 months, the infant was diagnosed as epileptic. Cardiologic examinations (ECG, echocardiogram, Holter monitor evaluation) and chest X-Ray were normal.

The autopsy revealed a well developed and well nourished 13month-old white male. All internal organs were in their normal anatomic relationship. The heart, in the fresh state, weighed 45 grams. Upon sectioning, the myocardium was reddish. Atrio-ventricular and semilunar valves were normal. The right coronary artery arose from the left sinus of Valsalva associated to a tunneled passage of the posterior interventricular coronary artery. The first section of this artery presented a take-off and a passage between the aortic and pulmonary root. The coronary circuit was dominant to the right.

The histological examination of the cardiac tissues revealed diffused and biventricular myocytolisis in contractile subendocardial bands characterized by altered, eosinophilic, hypercontracted myofiber. In many areas the myofiber appeared fragmented into irregular, partially acidophilic transverse bands. The immunohistochemical exams, effected on cardiac fragments, resulted positive to the anti - desmin and antiactin antibodies, and negative to the anti C5 antibodies. Examination of the other organs were unremarkable except for pulmonary edema and polyvisceral stasis.

Case 2: A 22-year-old white male collapsed and died while exercising in a swimming pool; prompt medical assistance and attempted resuscitation were unsuccessful. At the age of 21, he was hospitalized in a Neurological Clinic, where instrumental and clinical data suggested the diagnosis of Friedreich's ataxia. The subject underwent molecular genetic analysis for the FA gene that revealed neither expansion nor point mutation of the FA gene. One year later, he was hospitalized at another Neurological Clinic where a general examination showed kyphoskoliosis, pes cavus, and a neurologic examination showed nystagmus, hypotonus, distal hypotrophy of the arms and legs, ataxia, areflexia, abnormalities in superficial and deep sensations. Ncv and EMG examinations were compatible with a severe axonal-myelinic sensorymotor neuropathy, while vitamin E, B12, folic acid, antigliadin antibodies, hexosaminidase, transferring isoforms, lactate and pyruvate were all normal. The diagnosis was spinocerebellar heredodegeneration, Friedreich's type. A treatment with idebenone, CoQ100 and Vitamin E associated with physiotherapy was suggested.

At autopsy the body was that of a well-developed young adult. Skin, ostia, oral and scleral mucosae were normal. On evisceration the heart had a normal intrathoracic position with the following diameters: longitudinal 11 cm, transversal 13 cm and antero-posterior 7 cm; the weight was 475 gr. On opening, the ventricular chamber was 30 mm wide, the wall and the inter-ventricular septum measured 28 mm each. The endocardium was white, smooth and bright, no trombi or vegetations were detected. Atrio-ventricular and semilunar valves were normal. The right coronary artery normally arose from the right ostium. In the left sinus of Valsalva two distinct ostia were detected instead of the left coronary artery ostium. The diameters of the two ostia measured 1,5 and 2,5 mm respectively and were separated by a septum that divided the stem determining a separate origin of the two left coronaries. Incannulation and a careful dissection demonstrated that the larger ostium was one of the circumflex branch, the smaller one the ostium of the anterior branch.

The histological examination of the sections revealed diffuse interstitial fibrosis due to the presence of thin fibrous septa dividing the muscle cells. Miocytes showed fragmentation of the fibers, nuclear enlargement, sometimes pyknosis and cytoplasmatic vacuolization. A section taken from the interventricular septum showed hemorrhagic infiltration of the wall with single erythrocytes between the single myocites and small blood extravasations. Examination of the other organs were unremarkable except for pulmonary edema and polyvisceral stasis.

The autoptic findings and the histological studies effected lead us to conclude that these are both cases of sudden cardiac death in subjects affected by coronary anomalies.

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In the first case death was caused by cardiac arrhythmia sustained by myocardial hypoxia induced by an anomalous origin of the right coronary artery from the left Valsalva sinus.

In the second case death was caused by cardiac arrhythmia, sustained by myocardial hypoxia induced by an anomaly of the left Valsalva sinus, divided into two distinct ostium: one in the anterior intraventricular coronary and one in the circumflexed associated with myocardial hypertrophy, in subjects affected by Friedreich's ataxia.

Congenital coronary anomalies constitute a statistical incidence of 0,3-0,8% and represent 0,1-2% of all congenital cardiac conditions worldwide. If we consider the anomaly originating of the right coronary artery from the left Valsalva sinus, as revealed in one of the two cases examined, the prevalence from autoptic studies is reduced to 0.026%.

Congenital anomalies of the coronary arteries present great difficulties in diagnosis because these diseases can be absolutely asymptomatic and, although rarely, can manifest themselves with syncopal episodes or with a fading symptomatology leading to heart failure. However, the prognosis is influenced by the seriously of coronary anomaly.

The anomalous origin of the right coronary artery from the left Valsalva sinus has long been considered a mostly benign disease and only in 1982 three cases of sudden death are described whose cause depends on this type of congenital alteration.

In literature, the stenosis or coronary take-off in the initial tract are interpreted as causing ischemia and sudden death.

The origin of the right coronary artery from the left sinus may be an incidental observation during autopsy. Ischemia is usually precipitated by strenuous, prolonged effort, and this explains why a basal ECG or even a stress test ECG may be negative. Syncopal episodes are the only prodromal symptoms. Repetitive ischemic episodes may cause patchy myocardial necrosis and fibrosis as well as ventricular hypertrophy, which eventually can elicit arrhythmias because of the malignant combination of acute and chronic substrates. This may explain why sudden death, associated with an anomalous origin of a coronary artery from the wrong sinus, may occur in adults even though the anomaly has been present since birth.

An anomalous origin of the left circumflex artery from the left coronary sinus itself with a separate ostium, has also been described in victims of unexpected arrhythmic sudden death. This anomaly was considered a benign condition until cases were reported, both clinically and pathologically, with evidence of myocardial ischemia in the absence of obstructive coronary atherosclerosis or causes other than the malformation itself.

It should be noted that in cases of coronary anomalies sudden death, in children and young adults, often occurs during or following physical exertion. In the second case reported death occurred during physical activity and the anomaly of the left Valsalva sinus, divided into two distinct ostium: one for the anterior interventricular coronary and one for the circumflexed, was associated with cardiac hypertrophy, a pathology present very frequently in subjects affected with Friedreich's ataxia.

Sudden Death, Anomalous Origin Coronary Artery, Contraction Band Necrosis

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