G121 Aneurysms of the Coronary Arteries and Sudden Death: Three Case Reports

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After attending this presentation, attendees will be able to better understand the main pathological features and pathogenesis of Coronary Artery Aneurysms (CAAs) in atherosclerotic—one case, and Kawasaki Disease (KD)—2 cases.

This presentation will impact the forensic science community by increasing the awareness that CAAs bear a significant risk of sudden cardiac death and that their development depends on transmural vascular inflammation.

CAAs are rare with an incidence of 0.02% to 0.04% in the general population. They are most commonly associated with the male gender and hyperlipidaemia. The proximal right coronary artery seems to be predominantly altered. About one-fourth of these patients have multiple coronary aneurysms. CAAs most commonly develop secondary to atherosclerotic coronary artery disease. Some CAAs are described as congenital and are linked to KD. Other circumstances are rather exceptional (trauma and dissection, infectious, scleroderma, connective tissue disorders such Marfan or Ehlers-Danlos syndromes and lupus, neurofibromatosis, periartheritis nodosa, and Takayasu's arteritis). Intraluminal (parietal or occlusive) acute thrombosis is found in 75% of the aneurysms with possible development of myocardial ischaemia and Sudden Death (SD). SD may also occur subsequently after rupture of CAAs and cardiac tamponade.

The first case regards a 3-month-old Italian male baby who, three days after the compulsory vaccination and antipyretic administration to calm the feverish condition, gradually showed eyelid conjunctival and lip erythema, diffuse cutaneous exanthema, and pharingo-tonsillitis associated to staring episodes. At admission, his temperature was 39°C and the hemato-chemical data were: erythrocytes $3.51x10^6$/uL; leukocytes $9.92x10^3$/uL; platelets $626x10^3$/uL; alcaline phosphatase 334 IU/l, PCR 7.85mg/dl; ALT 49IU/l. An echocardiogram showed a diffused hyperechogenic right coronary, dilated in the proximal tract with a 4mm aneurismal margin; hyperechogenic left coronary, dilated at the level of the common trunk, with an aneurysmatic aspect at the level of the first tract of the anterior interventricular septum; moderate pericardial effusion; well preserved myocardial contractility. Death suddenly occurred nine days after admission. The autopsy revealed that death was due to cardiac tamponade subsequent to aneurysm rupture of the anterior descending coronary, the wall of which showed active transmural lymphomonocytic inflammation. The second case regards a 2-month-old male Indian infant. The infant showed signs of rhinitis and coughing associated with conjunctival hyperemia and allergic exanthema on the chest and arms. Laboratory analyses showed: leukocytosis ($15.370/mm^3$), elevated sedimentation rate; positive C-reactive protein, and thrombocytosis ($476.000/mm^3$). Klebsiella pneumoniae was isolated from the urine and an antibiotic therapy was started. Coughing, rhinitis, exanthema, and conjunctivitis progressively decreased until they disappeared, while laboratory tests showed an increase in phlogosis indices. Death came suddenly and unexpectedly on the seventh day of recovery. The autopsy revealed that death was due to cardiac tamponade subsequent to aneurysm rupture of the anterior descending coronary, the wall of which showed active transmural lymphomonocytic inflammation.

The third case concerns a 66-year-old man with a history of ischaemic heart disease and stroke cerebri, who entered the emergency area in a stupor state. The patient became comatose (Glasgow Coma Scale: 5) and the brain CT scan demonstrated a hyperdense right middle cerebral artery with a flattening of the cortical sulci along with signs of a suffering chronic hypoxia and after effects of an extensive malacic bilateral frontal area. The death occurred four hours after recovery. The autopsy showed a diffuse fibro-atheromasic thickening of the walls of the subepicardial coronary arteries which appeared focally aneurysmatic along with occlusive endoluminal thrombosis and chronic transmural flogosis.

The pathomechanism of CAAs remains a controversial topic of discussion. Weakened media of the coronary wall with a diminution of its elastic elements in areas of severe atherosclerotic plaques, and intraluminal pressure against the defective vessel wall, allow the vessel to dilate progressively. Not only does the chronic transmural inflammation destroy the media which represents the pathomechanism of CAAs in KD, but also makes an analogous possible pathway in atherosclerotic aneurysms. Our cases show that CAAs are an independent predictor of mortality because their development depends on the possible transmural extent of intimal (in atherosclerotic disease) and adventitial in KD inflammation. MRI represents the best diagnostic tool for identifying this rare pathological evolution.

Coronary Aneurysms, Atheroscler Disease, Kawasaki Disease

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