

Some practical cases of SCD with alleged medical negligence

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Highlights for Cardiac Cases. Stephen Cohle MD

1. Cystic Tumour of the AV Node

- Cause complete heart block
- Arise from endoderm
- Sudden death can occur despite pacemaker placement
- Can grow due to glandular secretions

2. Arrhythmogenic Cardiomyopathy (Right ventricular cardiomyopathy)

- Fibro-fatty replacement of the right and/or the left ventricle
- Desmosomal protein mutations
- Sudden death can occur in phenotypically normal hearts
- Sudden death occurs in athletes due to long term strenuous activity

3. Histiocytoid Cardiomyopathy

- Gross: raised endocardial plaques
- Pale, granular histiocytoid cells are mainly endocardial but can be found throughout the myocardium
- Cells are filled with mitochondria
- Can present with seizures, which can be misinterpreted as being of CNS origin

4. Left ventricular non-compaction

- Caused by fetal type myocardium which persists into adult life
- Trabecular/compact myocardium 2:1
- Classic triad: arrhythmias, heart failure, emboli

Updating in post-mortem imaging for SCD

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Abstract

Radiological examination has been used in forensic and in clinical pathology in specific situations of Sudden Death for many years, as an additional diagnostic tool, for documentation and for research. In Sudden Cardiac Death (SCD) cases, traditional plain x-rays images and Postmortem Computed Tomography (PMCT) are helpful to estimate the heart size and to visualize haemopericardium, calcified plaques and valves, as well as to identify and locate cardiovascular devices.

Angiographic methods are useful to provide a detailed view of the distribution pattern of coronary arteries and to analyse them, especially regarding the extent and location of arterial stenosis and obstruction. In post-surgical cases, it allows verifying patency of stents and bypassing grafts and may guide further sampling for histological examination [1-3].

Whole body imaging PMCT and Postmortem Magnetic Resonance Imaging (PMMR), associated or not with angiography methods, have been increasingly used during the last years, especially in forensic practice. Some religious groups have objections to invasive autopsy and demand for an alternative. Therefore, minimally invasive autopsy techniques have increased. The advantages and disadvantages of various radiological methods for whole body imaging are related to logistic and financial questions as well as those concerning the image quality [4-7]. Today, these methods are mostly seen as complementary techniques to standard autopsy, as they are said to be still less accurate than the reference standard for natural deaths [4, 6, 8-10]. Also they do not replace the histological examination.

One problem for investigating SCD by imaging methods is that the most widespread modern imaging technique, the PMCT, has a very limited application in cases of natural death as its accuracy for detecting cardiovascular findings is low. Only in a few cases related to massive hemorrhagic events, such as acute intracerebral hemorrhage, cardiac tamponade or ruptured aortic aneurysm, PMCT could potentially explain the death [4, 11]. PMCT-angiography however, allows the evaluation of vessels including coronary arteries [3, 12, 13], gives information concerning the origin of a hemopericardium (rupture of infarcted myocardium versus aortic dissection) and may also guide further sampling for histological examination [1, 3, 12, 13]. This is why PMCT-Angiography is today the imaging method of choice for examining cases of SCD.

PMMR is available only in a few academic centres and practiced to investigate the soft tissues such as the myocardium. Also PMMR-angiography is today in development [14-17].

Detection of myocardial ischemic injury has been reported, but images are susceptible to be affected by post-mortem changes and further works are necessary to increase the understanding of radiological aspects of the myocardium [18, 19].

This presentation will give an update about existing imaging techniques and explain their advantages and limitations. It will show the participants the application of the methods by using illustrating case examples. It will also discuss the role of such imaging techniques in today's postmortem investigation of SCD.

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Role of Forensic Pathologist in the Investigation of a sudden cardiac death (SCD)

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Abstract

The sudden death (SD) of a person, usually young, in apparently good health, although infrequent is a very dramatic event with social and clinical implications and great concern for bereaved families.

According to the commonly accepted criteria, SD is defined in witnesses' cases as a natural death that occurs within 6 hours of the beginning of symptoms in an apparently healthy subject or one whose disease was not so severe that a fatal outcome would have been expected. In cases of non-witnessed death, the definition requires that the deceased was last seen alive and functioning normally 24 h before being found dead.

Due to the rapid and unexpected onset of the death, which appears in an apparently healthy person, a forensic autopsy is usually performed in the majority of countries with the aim to exclude a violent mechanism and to determine the cause and manner of death. In this context, autopsy represents the first and last opportunity to make a proper diagnosis and thereby a valid mortality registration. In this regard, forensic registries are a reliable source to assess the impact of this problem in a given population. On the other hand, autopsy plays a pivotal role in the identification of families at risk and the pathologists job is essential in the counselling to refer first degree family members to cardiological screening and additionally to perform post-mortem genetic testing.

The underlying cause of sudden death is most frequently cardiovascular with coronary atherosclerotic disease as the leading cause of death in victims ≥ 35 years of age whereas in those < 35 years the leading cause is sudden unexplained death. Progresses made in the fields of molecular biology and human genetics have identified the genetic origin of many cardiac diseases, which can lead to both, structural (e.g. HCM, ARVC/D. DCM) and non-structural or arrhythmogenic abnormalities (e.g. LQT syndrome, Brugada syndrome) and result in sudden cardiac death (SCD). Autopsy-negative SCD is most often thought to be the consequence of sudden arrhythmic death syndrome ("mors sine material") and post-mortem genetic testing (molecular autopsy) is recommended.

In our department, autopsies are performed following the Recommendations on the Harmonization of Medico-Legal Autopsy Rules produced by the Committee of Ministers of the Council of Europe and the guidelines for the autopsy investigation of SCD from the Association for European Cardiovascular Pathology. Specifically, a protocol is followed which included: clinical antecedents of the case, death scene investigation, body height and weight, waist circumference, and complete macroscopic autopsy with histology of all organs.

The study protocol should include not only gross examination of the heart and histological sampling, but toxicology, microbiology, biochemistry and molecular investigation.

In summary, the role of a forensic autopsy in a SD is to establish:

- If the death is natural or unnatural (violent).
- The possibility that a toxic or drug of abuse may have contributed to the death.
- If the death is from a cardiac or extra-cardiac origin.
- The nature of the cardiac disease and whether the mechanism was arrhythmic (electrical SCD) or mechanical.
- If the cardiac condition causing SD may be inherited, thus requiring the cardiac assessment of first-degree relatives and to perform post-mortem genetic screening in the proband.
- A multidisciplinary approach in a SCD is essential with close contact between pathologists, cardiologists and geneticists.

In this presentation, eminently practical, we'll update the knowledge in sudden unexpected death related to cardiovascular pathology and will improve the skills in the diagnosis and approach of SCD in the setting of forensic medicine and pathology.

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