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Sudden death related to anomalous origin of coronary artery and coexisting fenestrated membrane of the sinus coronarius

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ABSTRACT

Coronary artery anomalies that may be an isolated defect or part of complex congenital malformations of the heart are also often associated with a high risk of sudden death. A 19-year-old woman lost consciousness in the night. She was taken to hospital where she was treated. However, she died on the same day. The prosecutor considered the death suspicious, and the victim was taken to the morgue department for further examination and autopsy. On macroscopical examination, both coronary arteries originated from the left aortic sinus with the left one being dominant. The orifice of the sinus coronarius in the right atria was covered with a fenestrated membrane. Demonstration of coronary artery pathologies and associated anomalies in autopsies is vital for the elucidation of sudden death cases related to these lesions and for the development of new treatment approaches. The purposes of this case report are to characterise and identify markers of the coronary artery anomalies and to highlight their medicolegal importance in sudden death cases.

Keywords: anomalous origin of coronary artery, aortic sinus, coronary artery, sinus coronarius, sudden death

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INTRODUCTION

Coronary artery anomalies that may be an isolated defect or part of complex congenital malformations of the heart are also often associated with high risk of sudden death.[1] Right or left coronary arteries originating from the wrong sinus of Valsalva cause non-specific symptoms. However, they are detected rarely on angiographies and autopsy series.[1,2] There are reports of sudden death in athletes who, on autopsy, are diagnosed as having coronary artery origin anomalies.[2] Episodes are reported to occur after exertion,[2,3] but deaths without any symptoms have also been encountered.[4] Recognition of coronary artery pathologies in autopsies is mandatory for the explanation of sudden death cases related to these lesions and for the improvement of new therapeutic approaches. The purposes of this case report are to characterise and identify markers of the coronary artery anomalies and to highlight their medicolegal importance in sudden death cases.

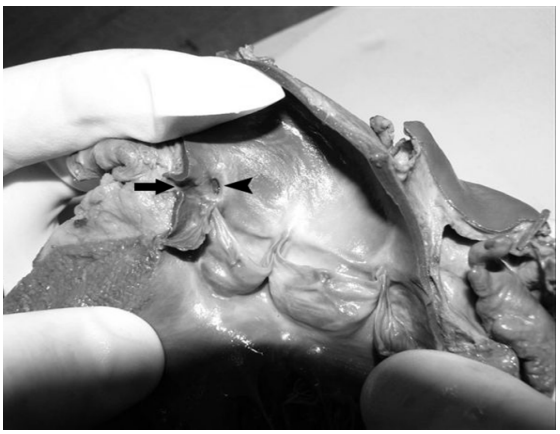

Fig. 1 Specimen photograph shows coronary arteries originating from the left sinus aorticus, left coronary artery (arrow) and right coronary artery in the upper part of the aortic sinus (arrowhead).

CASE REPORT

According to the document of death, the patient, a 19-year-old woman, lost consciousness in the night. She was taken to a hospital emergency department where she was treated. However, she died on the same day. The prosecutor considered the death suspicious and the victim was taken to the morgue department for further examination and autopsy. Her family members stated that she had had occasional fainting episodes, but as she had never been medically evaluated, there was no available information about character, frequency, and recovery of the episodes. The case was a 19-

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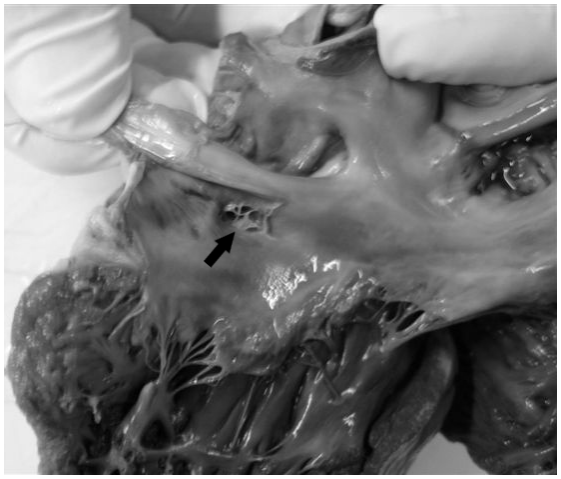

Fig. 2 Specimen photograph shows a fenestrated membrane at the sinus coronarius (arrow).

year-old cadaver; 160 cm tall and weighed 50 kg. On gross physical examination, there were defibrillator-related burn areas on the chest wall and needle puncture sites on the back of the right hand and the cubital fossa. Autopsy macroscopic examination revealed petechial subpleural haemorrhages on interlobar surfaces of both lungs, while microscopic investigation detected oedema and congestion. The brain appeared normal on histopathological examination.

The heart weighed 250 g. There were petechial areas behind the atria, and the aortic arch and valves appeared normal. Both coronary arteries originated from the left aortic sinus (Fig. 1), and the left one was dominant. The right coronary artery originated from the upper part of the aortic sinus and crossed superficially between the aorta and the pulmonary artery. The ostium had the shape of a slit. The orifice of the sinus coronarius at the right atria was covered with a fenestrated membrane (Fig. 2), but the coronary sinus was not dilated as a result of obstruction. There was a 3 cm × 1 cm subendocardial bleeding area on the upper part of the left ventricular septum. Six sections from the heart were evaluated. On histopathological examination, there was no evidence of acute or chronic ischaemia. Toxicological systematic studies of the organ specimens revealed no special finding. Death was reported as sudden natural death due to anomalous origin of coronary artery.

DISCUSSION

Coronary artery anomalies that entail a risk of sudden death – right or left coronary arteries originating from the wrong sinus of Valsalva – cause non-specific symptoms and are rarely detected on angiographies.[1,2] A left coronary artery originating from the right sinus Valsalva is reportedly more frequent; our patient had the rarer variant – a right coronary artery originating from the left sinus of Valsalva.[1-3,5-7] In an angiographical study from our country, Ayalp et al reported a frequency of 0.05% for the right coronary artery originating from the left sinus of Valsalva.[8] Some authors suggest that these may be associated with bicuspid aorta.[7] The structure encountered at the sinus coronarius is referred to as the Chiari fenestration in literature (frequency 1.5%–3%). It has been reported to be associated with asymptomatic foramen ovale, thromboembolism, atrial aneurysms and arrhythmias,[9] but association with coronary artery anomalies was not mentioned before in literature.

Chiari fenestration represents a persistence of the right sinus venosus valve. Normally, the right sinus venosus valve regresses and the caudal portion develops into the Eustachian and Thebesian valves. Any failure in the regression process may result in remnants of the right sinus venosus valve as a simple muscle bar, a Chiari-network or a fenestrated membrane.[10] The orifice of the coronary artery may be slit-like; during exertion, the coronary artery is compressed between the aorta and the pulmonary artery, and this results in impaired oxygenation and myocardial ischaemia that may result in sudden death.[1,3,6] As was the case in our patient, they may give no symptoms until the lethal episode.[4] Studies on athletes have shown that the deaths in patients with coronary artery origin anomalies usually occur during or after exertion.[1-3] Bunai et al reported that the right coronary artery originating from the left sinus of Valsalva is a risk factor for sudden death associated with exercise but may also cause sudden death unrelated to exertion.[6]

The literature suggests that the risk of death is higher in patients younger than 30 years and comparatively lower at more advanced ages.[3] Syncope attacks (present in our patient) possibly associated with cardiac ischaemia may be observed,[1,2] and chest pain and palpitation have also been reported.[6,8] Recognition of coronary artery origin anomalies in autopsies is mandatory for the explanation of sudden death cases related to these lesions and also for the improvement of new early therapeutic approaches.[1,3] The coronary anomaly can be corrected successfully by unroofing procedures, where manipulation of the commissure can be avoided by creation of a neo-ostia without extensive unroofing of the intramural segment or manipulation of the intercoronary commissure, which may avoid aortic valve malfunction.[5] Autopsy procedures done for determining the coronary artery anomalies related to sudden death are of great importance for the final medicolegal investigation of the cases.

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