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Death due to coronary artery anomaly with coexistence of Chiari network

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SUMMARY

Coronary artery anomalies rarely detected in autopsy series and angiograms can be a component of complex malformations, besides, can be also associated with sudden cardiac death. Presented case was 22-year-old male, who had suddenly fainted during a football match played on artificial turf, he was transferred into the hospital, however had died during intensive care therapy. He had been evaluated by local prosecutor, and sent to our center for autopsy. At autopsy, internal macroscopic examination revealed absence of the right coronary artery. A total of two coronary artery ostia were observed. One of them originated from the left aortic sinus, and the other one stemmed from 8 mm above the sinotubular line. Besides, Chiari network formation was seen in the right atrium. This case with coronary artery anomaly associated with formation of Chiari network was discussed from the perspective of forensic medicine in the light of the literature information.

Keywords: coronary artery – Chiari network – sudden death – autopsy

Náhlá smrt při anomálii věnčité tepny asociované s přítomností rete Chiari

SOUHRN

Anomálie věnčitých tepen jsou vzácně odhaleny při pitvě srdce anebo při koronarografii; mohou být součástí komplexních malformací, popř. mohou vést k náhlé srdeční smrti. V našem příspěvku uvádíme případ 22letého muže, který náhle zkolaboval při fotbalovém zápase odehrávaném na umělém trávníku. Muž krátce po převozu do nemocnice, i přes intenzívní resuscitační péči, zemřel. Rozhodnutím státního zástupce byla nařízena pitva a tělo muže bylo odesláno k pitvě na naše pracoviště. Dle sdělení příbuzných nikdo z rodiny netrpěl onemocněním srdce. Při vnitřní prohlídce byla zjištěna nepřítomnost pravé věnčité tepny. V oblasti sinu aorty byla identifikována dvě koronární ústí. První ústí bylo patrné v levém koronárním sinu. Kmen věnčité tepny z něj vycházející se dělil na dvě větve; první větev kopírovala průběh sestupného ramene levé věnčité tepny, druhá sledovala průběh pravé věnčité tepny. Druhé koronární ústí bylo v levé části vzestupné aorty 8 mm nad sinotubulární čárou a mělo průměr 7 mm, věnčitá tepna z něj odstupovala pod úhlem 45 stupňů. V pravé síni byla navíc identifikována Chiariho síťka. Chiariho síťku poprvé popsal v roce 1897 Hans Chiari. V příspěvku je v soudnělékařském kontextu a ve světle současné literatury popsán případ náhlé srdeční smrti s absencí pravé věnčité tepny a přítomností Chiariho síťky v pravé síni.

Klíčová slova: koronární tepna – rete Chiari – náhlá smrt – pitva

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Coronary artery anomalies can be a component of complex congenital malformations or an isolated defect. Besides, they are closely associated with cases of sudden death, but they are rarely detected in autopsy series of coronary artery anomalies, and on angiograms (1,2). Anomalous origins of coronary arteries have been associated with sudden deaths in young athletes (2). In the literature, prevalence of Chiari network has been reported as 1.5-2%, and it is related to thromboembolic events, and cardiac arrhythmias (3,4). In the literature, its incidence in autopsy series has been reported as 10.52 percent (5). Generally Chiari network has not any clinical significance (3,4). Nowadays, with widespread use of echocardiographic examinations in hospitals, and rapid development of this imaging technique, this congenital remnant can be diagnosed (3-6). Forensic medicine specialists should recognize these lesions in autopsies in order to explain the association between these lesions, and sudden deaths, and to develop new therapeutic approaches. Herein, we discussed from the perspective of forensic medicine and in the light of literature findings, a sudden death of an individual who had a coronary artery anomaly associated with formation of Chiari network, which had an asymptomatic progression till death of the patient.

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CASE REPORT

On 4 March 2013, a 22-year-old young man had suddenly fainted at 11.00 a.m., and fallen on the ground while running during a football match played on artificial turf, and after first aid provided by the emergency ambulance personnel, he had been transferred to a hospital, and died during his treatment in the intensive care unit. He had been evaluated as a suspect case of death, and transferred to our center for autopsy. As indicated by his family, any type of cardiac disease was not present in family members, and the deceased. It was also learnt that he had completed his military service, and had not any cardiac

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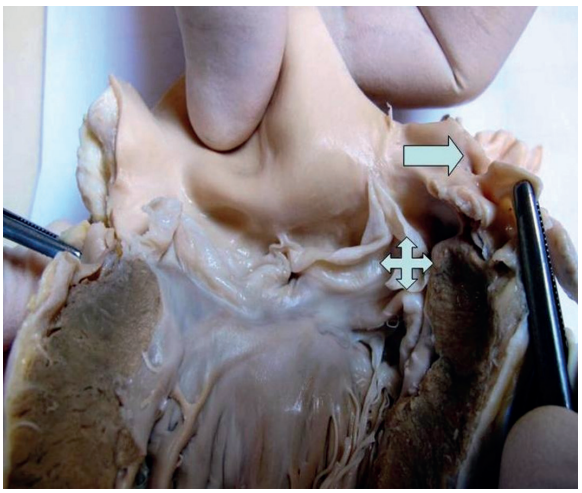


Fig. 1. A four-way arrow pointing the ostium of the left coronary artery, an arrow pointing to the ostium of the ectopic coronary artery.

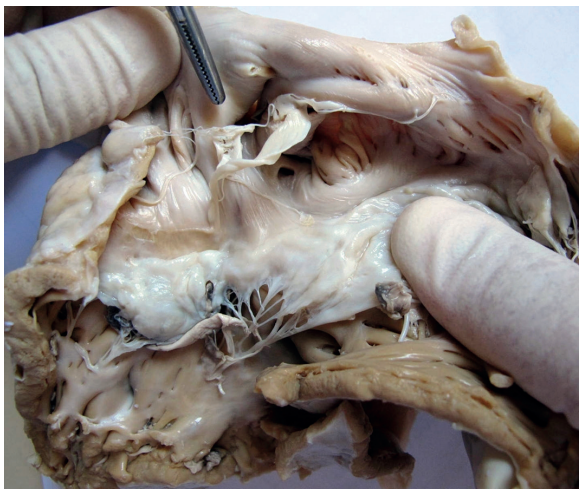


Fig. 2. Chiari network in the right atrium.

complaints before, during, and after completion of his military service. At autopsy, any sign of trauma was not detected during external physical examination of the corpse except for extremely cachectic body, tracheostomy, and decubitus wounds. Internal examination was performed. His heart weighed 306 g. Right coronary artery was not detected. A total of two coronary ostia were observed. One of the coronary arteries originated from the left aortic sinus, the other with a diameter of 7 mm, stemmed with a 45 degree angle from the upper left part of the ascending aortic wall 8 mm above the sinotubular line (Fig. 1). Coronary artery originating from the left aortic sinus divided into two branches. One of these branches coursed along the descending coronary artery, and the other one followed the track of the right coronary artery. Ectopic coronary artery advanced along the course of circumflex coronary artery. Besides Chiari network formation was observed in the right atrium (Fig. 2). On the surfaces of both lungs, anthracotic pigmentation, and petechial hemorrhagic foci were seen, and sections obtained demonstrated the presence of edema, and yellow-green mucoid material. Inside the abdominal cavity, accumulation of whitish yellow fluid was also noted. Any histopathological abnormality was not detected in other organs. Analysis of the medical records of the deceased revealed that he had been connected to mechanic ventilator for nearly 4 days in the intensive care unit (ICU) with the diagnosis of cardiac arrest. He had been under supportive treatment in ICU, and undergone tracheostomy, and jejunostomy operations. However his clinical manifestations did not change, and cardiac arrest recurred which necessitated employment of resuscitative measures. Despite all these resuscitative efforts his condition had not improved and attendant physicians eventually had admitted his demise. Biochemical analysis of his blood, and urine samples had not disclosed presence of any culprit analyte. Cause of death of this patient with detected Chiari network formation inside the right atrium was recorded in his death certificate as sudden death caused by anomalous origin of coronary artery.

DISCUSSION

Anomalous origins of coronary arteries have attracted forensic medicine specialist as a condition that unfavourably contributes to the risk of sudden death (1,2,7-10). In angiography series, the incidence of coronary artery anomalies has been reported to vary between 0.29, and 3.35 percent (11-14). In the literature, ectopic, and abnormally high coronary artery ostium has been defined as an ostium localized 5 (7,8) or 10 mm above the sinotubular line (9,10). Chiari network formation was originally described by Hans Chiariin 1897 as fibrillar net in the right atrium (4). Normally, right-side valve of the sinus venosus regresses during the embryologic development. If any abnormality occurs during this process, it persists as a remnant on eustachian, thebessian valves, and crista terminalis. This mobile reticular network is called Chiari network (6,16). In the literature, the prevalence of Chiari network formation has been reported as 1.5-2% which is diagnosed during echocardiographic examination (3-4,16). We previously presented a case report on a deceased patient with Chiari network associated with anomalous coronary artery. As we have mentioned before this unusual association has not been cited in the literature, so far (15). As far as we know, our present case is the second similar case mentioned in the literature. Twenty percent of coronary artery anomalies induce arrhythmias, syncope, myocardial infarction, and sudden deaths, while 80 % of them have a benign course (13). Left circumflex, and left descending coronary artery anomalies originating from the ostia apart from the left valsalva sinus are the most frequently seen coronary artery anomalies (11,13,14,17). These anomalies are not etiological factors for hemodynamic disorders, and they are evaluated as benign coronary artery anomalies. Besides, it is difficult to detect shorter left coronary arteries or absence of them during angiographic examinations (13). Anomalous coronary artery with a high ectopic insertion can decrease myocardial perfusion; however this condition is also related to angulations between coronary artery ostium, and aortic wall (8). Angulation at the coronary artery ostium functions as a valve. During exercise, the velocity of blood flow within the ectopic coronary artery anomalies with a high ectopic ostia increases, and upper part of the ostial wall is retracted to the inferolateral direction, while its lower part is pulled upwards leading to ostial occlusion (7,10,18). In our case, histopathological examination of the cardiac tissue samples did not reveal any sign of myocardial ischemia, apart from congestion, and enlargement of cytoplasm of myocytes. Coronary artery anomalies are very closely associated with sudden deaths (1,2), and most of them lead an asymptomatic course (13).

Forensic medicine specialists should recognize relevant lesions very well during their autopsy practices in order to aid in the detection of sudden cardiac deaths, classification of coro-

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nary artery anomalies, and development of new management strategies. In the future, greater amount of data retrieved from large-scale studies will clarify the association between Chiari network, and coronary artery anomalies.

CONFLICT OF INTEREST
The authors declare that there is no conflict of interest regarding the publication of this paper.

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ZPRÁVA ZE SJEZDU

III. Kongres traumatologie a muskuloskeletální radiologie

Ve dnech 16. – 17. dubna 2015 se v konferenčních prostorách hotelu Myslivna v Brně konal III. Kongres traumatologie a muskuloskeletální radiologie. Akce byla rozdělena celkem do 7 sekcí soustřeďujících se většinou na problematiku páteře a kolenního kloubu. I když akci pořádala Radiologická klinika Lékařské fakulty MU Brno, část týkající se traumatologie přinesla i některé zajímavosti využitelné pro praxi v soudním lékařství. Zejména práce „**Úrazy horní krční páteře**" autorů Ryba L. et al. z Ortopedické kliniky Fakultní nemocnice Brno pojednávající nejen o diagnostice a terapii, ale popisující velice pečlivě a podrobně mechanizmus vzniku jednotlivých typů úrazů včetně schematických znázornění, by zasluhovala prezentaci i v prostředí určeném soudnělékařské veřejnosti. To jenom dokazuje multioborovost soudního lékařství a prospěšnost jeho vzájemné provázanosti s ostatními obory medicíny.

Miroslav Hirt, Brno

