Truncus arteriosus communis with survival to the age of 46 years: case report

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SUMMARY

Truncus arteriosus communis is an uncommon congenital cardiovascular malformation characterized by a single arterial trunk that arises from the base of the heart and gives rise to the coronary, pulmonary and systemic arteries. The prognosis in truncus arteriosus is very poor without surgical correction. The median age at death without surgery ranges from 2 weeks to 3 months, with 85 % mortality by age 1 year. The authors report the autopsy findings of a 46 year old man with truncus arteriosus communis without surgical intervention who died at the hospital shortly after admission.

Keywords: truncus arteriosus communis – heart failure – autopsy findings

Truncus arteriosus communis s prežívaním 46 rokov: kazuistika

SÚHRN

Súdny lekár sa častokrát stretáva s prípadmi náhleho a neočakávaného úmrtia v zdravotníckom zariadení, u ktorých nie sú známe anamnestické údaje o ochoreniach, prípadne tieto údaje sú nedostatočné. Pitva často odhalí nálezy, ktoré sú neobvyklé. Medzi takéto raritné nálezy patria aj rôzne vývinové chyby kardiovaskulárneho systému. Autori prezentujú pitevný nález u 46-ročného muža s truncus arteriosus communis bez chirurgickej intervencie, ktorý zomrel krátko po prevoze do nemocnice za známok kardiorespiračného zlyhania. Truncus arteriosus communis je zriedkavá vrodená kardiovaskulárna anomália, pri ktorej výtoková časť pravej aj ľavej komory ústi priamo do spoločného arteriálneho kmeňa s jednou súpravou chlopní, ktorý obstaráva koronárnu, pľúcnu a systémovú cirkuláciu. Táto zriedkavá anomália je výsledkom zlyhania septácie primitívneho arteriálneho trunku počas embryonálneho života. Jednotlivé typy sú definované na základe miesta odstupu pľúcnych tepien z arteriálneho kmeňa. Celosvetovo predstavuje asi 1-2 % všetkých vrodených vývinových chýb srdca. Incidencia je 5-15 prípadov na 100 000 živonarodených detí. Ide o malformáciu, ktorá bez chirurgickej liečby má veľmi zlú prognózu. Bez liečby je táto kardiovaskulárna anomália zvyčajne fatálna a len asi 15 % jedincov prežije 1 rok života. Celkom výnimočne sa môžu títo jedinci dožiť vyššieho veku aj bez chirurgickej intervencie ako to bolo aj v prezentovanom prípade. Operačné riešenie truncus arteriosus communis nebolo vykonané pre technickú nedostupnosť na Slovensku v čase diagnostikovania anomálie u tohto muža.

Kľúčové slová: truncus arteriosus communis – srdcové zlyhanie – pitevný nález

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Forensic pathologists often investigate deaths that occur suddenly and unexpectedly shortly after hospital admission. Obtaining information may be limited at this time, especially when the deceased medical history is unavailable. Autopsy may reveal unusual findings, such as rare cardiovascular malformations. Therefore, choosing proper dissection techniques to clarify the cause of death is essential.

Truncus arteriosus communis is an uncommon congenital cardiovascular anomaly characterized by a single arterial trunk that arises from the base of the heart and gives rise to the coronary, pulmonary and systemic arteries. A single semilunar valve is found in truncus arteriosus. The anomaly is thought to result from incomplete or failed septation of the embryonic truncus arteriosus. Truncus arteriosus represents 1-2 % of congenital heart defects in liveborn infants (1,2). It occurs in approximately 5-15 of 100 000 live births (2). The authors present the case of a man with truncus arteriosus communis with survival to the age of 46 years without any surgical correction.

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CASE REPORT

We report the case of a 46 years old man with mild intellectual disability and congenital cardiovascular malformation – truncus arteriosus communis. After the autopsy, we requested to view his complete medical records and learned that he had nine siblings. Two of his brothers died of an unspecified congenital heart disease. Initially he was diagnosed with tetralogy of Fallot in 1972 when he was 6 years old. In 1975 this diagnosis was modified to truncus arteriosus communis and the condition was deemed inoperable at that time. He overcame common childhood illnesses and was repeatedly hospitalized for bronchopneumonia and cystitis as a child.

He was treated for arterial hypertension and in 2010 had episodes of grand mal seizures with cerebral hypoxia. From July till December 2011 he was hospitalized four times at the Department of Pneumonology and Phtiseology and once at the Department of Anesthesiology and Intensive Medicine. The reason for his admission was recidiving hemoptysis and hemoptoe.