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CASE REPORT / OLGU SUNUMU

The heart with single ventricle detected by exhumation and forensic autopsy: A case report

Fethi kabir ve adli otopsi ile tanı konulan tek ventriküllü kalp: Bir olgu sunumu

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ABSTRACT

Single ventricle heart is a rare cardiac abnormality identified by just a single ventricle involving various functional and physiological defects. This abnormality may cause sudden cardiac death. In this case, we reported a 71 day infant died on the fifth day after discharged from hospital who was buried without legal procedures. Autopsy findings were single ventricle, tricuspid atresia and aortic coarctation, a large heart weighing 134 grams. Our aim by reviewing this case report is to present the legal burial procedures, the autopsy and the clinical features of the subject.

Key words: Single ventricle, autopsy, exhumation

ÖZET

Tek ventrikül kalp, tek ventrikülle beraber çeşitli fonksiyonel ve fizyolojik kusurları içeren nadir bir kardiyak anomalidir. Bu anomali ani kardiyak ölüme neden olabilir. Bu olguda biz hastaneden taburcu olduktan sonra beşinci günde ölen ve defin ruhsatı alınmadan gömülen 71 günlük bebeği sunduk. Otopsi bulgularında tek ventrikül, triküspid atrezisi ve aort koarktasyonu, 134 gram ağırlığında büyük bir kalp vardı. Bu olgu sunumu ile amacımız bu konuyla ilgili yasal gömme prosedürleri, otopsi ve klinik özellikleri sunmaktır.

Anahtar kelimeler: Tek ventrikül, otopsi, fethi kabir

INTRODUCTION

Single ventricle heart defect is a rare cardiac anomaly, which generates 1.33% of all cardiac malformations [1]. This cardiac anomaly has various functional and physiological defects [2-5] and may cause sudden death [4,6]. The objective of this study is to present the legal burial procedures, the autopsy and the clinical features of the subject that was exhumed 9 days after interment.

CASE REPORT

Morgue Expertise Department of Bursa Group Administration Medical Examiner's Office performed an autopsy on the subject following exhumation; the legal and medical documents, autopsy findings, histopathology and chemical analysis results of the subject were studied.

Subject was born weighing 3018 g, with cesarean section after 40 weeks of conception he was admitted to the newborn clinic due to right choanal atresia, short frenulum, undescended left testicle and congenital heart disease. Echocardiography showed tricuspid atresia, VSD, ASD, mid-heavy mitral insufficiency, aortic coarctation. During the follow-up, the subject was intubated and connected to a ventilator upon sepsis complication. Surgery was not the preference at early stage; after the mother was given instructions, the infant was discharged from hospital after 66 days of birth. The infant was died after five days later of discharged at home and buried unlawfully by the family; he was exhumed 9 days later and a legal autopsy was performed.

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FINDINGS

Subject was 3368 g, 56 cm tall, 71 day old male infant. In the autopsy, external physical examination showed green livor mortis on the abdominal region due to decomposition and some postmortem hypostasis at the back of the body blind sight to the head on supine position. Also rigor mortis had ended. Internal examination showed that the heart weighed 134 g and was large. Dissection of the heart showed single ventricle, tricuspid atresia and aortic coarctation (Figure 1). Histopathological examination showed hypertrophic changes in the cardiac muscles and edema in the lung slides. Chemical analysis was negative for toxic substance. It is reported that the cause of death of the infant was congenital cardiac anomaly.

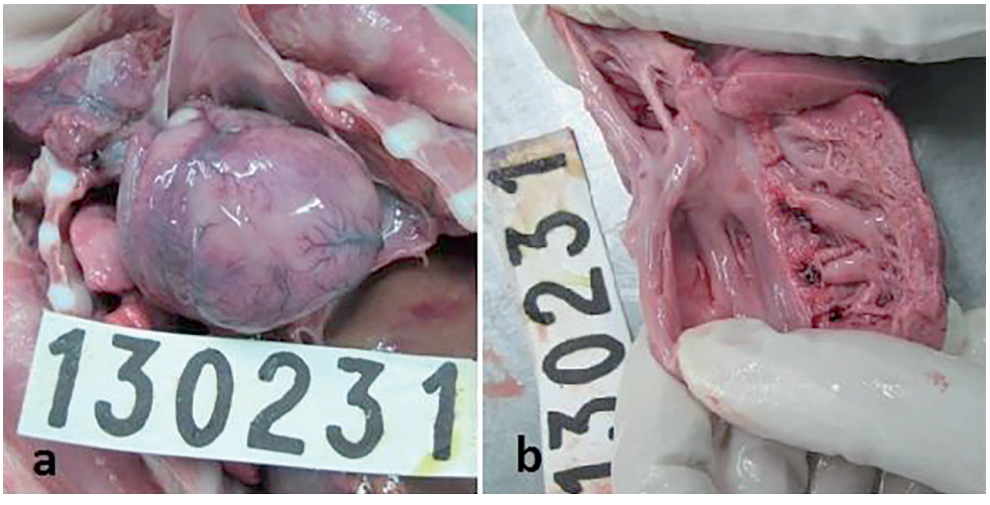

Figure 1: a. Macroscopic appearance of the heart. **b**. Single ventricle.

DISCUSSION

According to the Public Health Law in Turkey, burial of the dead is illegal unless interment permit is obtained and submitted. In our country interment permit can be issued by the government physicians, medical staff, the chief of military police stations and village headmen. Those who issue interment permit are not allowed to issue one if they suspect or conclude that the death was due to a contagious disease or an accident, before informing the relevant authority. In the case of being no physician, interment permit is issued for the deceased if in doubt of contagious disease that cause death and then relevant authority is informed [7]. Considering our case, upon his death at home, the subject who was concluded to have had congenital heart disease by post mortem studies was buried by his family without interment permit. We believe that the family did not know that they needed an interment permit to bury their baby, who already had a congenital heart disease.

There can be various cardiac anomalies and these can cause sudden cardiac death [8]. Single ventricle heart is one of this cardiac anomaly. Anatomically, patients with such defects have single ventricle. But mostly, functional single ventricle comprises a well-developed ventricle accompanied by a less developed ventricle [9]. Even though there are two ventricles, one being rudimentary, it is still called ‘‘functional single ventricle’’ since only one ventricle assumes the pumping function of the heart [10]. A well-developed ventricle’s morphology can be right ventricle manner with infero-posterior positioned rudimentary left ventricle (10-24%) or more commonly left ventricle manner with antero-superior positioned right ventricle (60-65%). In other words, left ventricle morphology is more common [4,9]. Sano et al, state that the right ventricle mannered subject’s ventricle wall thickness and mass is relatively less than left ventricle mannered subject’s ventricle wall thickness and mass [11].

Our investigation showed that the presented subject is the only case reported to have had single cardiac ventricle upon exhumation process. Usually these cases are diagnosed at newborn stage but Temizkan et al, reported a case diagnosed at the age of 16, presented at the age of 23 with medical abor-

tus indication during 1.5 months of into pregnancy and had a cerebral emboli a month later [12]. As it was presented here, it is also possible to see cases of medically proven illnesses with later diagnosis at the health institutions.

Clinical exams show subjects with single cardiac ventricle having cyanosis due to systemic arterial oxygen saturation decrease. These cases show decrease in injection fraction and seem to be candidate for pulmonary, aortic and atrioventricular valve regurgitation, arrhythmia and also infective endocarditis [4].

It is stated that consanguineous marriages have higher risk for congenital heart diseases (especially ASD, VSD, single ventricle and hypoplastic left heart) [13]. We do not have any information whether our subject's parents were blood-bond related.

In case of nonexistent pulmonary artery stenosis, usually patients who are not operated might have a large shunt from left to right with increased pulmonary blood circulation, advancing heart failure and besides intensive strain on the single ventricle that might lead to death. If there is pulmonary stenosis, then the pulmonary blood circulation is ascertained by the extent of the stenosis. The extent of the pulmonary blood circulation and the intensity level of stenosis will designate the time for surgical treatment. Occasionally, as a result of moderate pulmonary stenosis, it is seen that normal or a little elevated pulmonary pressure and sufficient pulmonary blood circulation are balanced [14].

Moons et al, states that as a result of the improvement in operational medicine, interventional and medical technology, life expectancy for children born with congenital heart disease have substantially increased and will continue to increase. Pediatric cardiologists and surgeons treatment options have considerably changed compared to the past. Hence, they state that new epidemiological researches are required for children born with congenital heart diseases in 21st century [15]. Prevalence studies of congenital anomalies are useful for determining the rate of anomalies, recording the changes by time and identifying the clues for etiology. It is also important for planning and evaluation of prenatal scanning for congenital anomalies, especially in high risk populations [16].

In conclusion, our society should be informed by individuals and institutions in order to avoid burials without a permit. In case of death, the deceased should not be buried without a permit. Besides, it is important to follow-up and examine the mother prior to pregnancy, as well as the baby during pregnancy in order to prevent congenital anomalies such as single ventricle. The disease should be detected, intensely followed-up and possible medical treatment is required; patient must be in close care and in some cases, surgery must be considered as an option.

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