

AN UNCOMMON CAUSE FOR A SUDDEN POSTOPERATIVE DEATH —A CASE REPORT—

Dayapala A & Fernando J
Teaching Hospital, Colombo North, Sri Lanka.

Introduction

When a sudden death occurs postoperatively after a routine surgical procedure for a non fatal condition, meticulous investigation is necessary to ascertain the cause of death as well as to support or refute medical negligence charges. This is more difficult in cases involving otherwise healthy young persons. In local set up, all concerned have to appreciate current practices in Sri Lankan hospitals and limitations in investigations. In such cases the Forensic Pathologist has to go into all the details in the history, investigations, the surgical procedure, possible complications, autopsy findings and especially histopathology. Sometimes it is necessary to review medical literature to address medico- legal issues credibly. This case report illustrates the importance of performing histological assessment of all organs including right ventricle and review of medical literature to educate ourselves of uncommon pathological entities and their effects.

History

Anotherwise healthy, 24- year- old unmarried female had a thyroid nodule. Fine Needle Aspiration Cytology revealed atypical cells and excision was recommended.

After getting informed written consent, investigations including routine blood tests, electrocardiogram (ECG) and Chest X-ray have been performed. They have not shown any abnormality.

After examining the patient and reviewing the results of routine investigations, the Consultant Anaesthetist decided that the patient was fit for general anaesthesia and surgery.

A near total thyroidectomy was performed and recovery from anaesthesia had been uneventful. As there were no Intensive Care Units beds available, she was sent to the ward.

Several minutes after being sent to the ward, she had vomited once. Immediately after that, she was found to be pulseless with unrecordable blood

pressure. Her breathing had become laboured and she developed sudden respiratory arrest.

She was rushed to the theater again with cardiac massage and ambu bag ventilation. ECG monitoring had showed a straight line. Intubation and vigorous resuscitative efforts has been performed by the anaesthetist in the theater without success.

Postmortem Examination

The body was average built without pallor, cyanosis, oedema or icterus. There was no bleeding from the ear, nose, throat, injection sites or operation site.

The endotracheal tube was in situ with inflated cuff. The operation site had two corrugated red rubber drains and was free from haematoma or surgical emphysema. The pleural cavity was free of haemopneumothorax or adhesions. There were no signs of aspiration into airways. Lungs were congested and oedematous but free of features of thromboembolisation.

The heart weighed 244g and valves, myocardial thickness and coronaries were unremarkable. But there was an ostium secundum atrial septal defect. (1cmx1cm)

Other organs were unremarkable to the naked eye.

All the organs were subjected to histological evaluation.

The right ventricular myocardium showed areas of severe fatty infiltration extending from pericardium to endocardium without inflammatory reaction or fibrosis. In some areas myocardial fibers were limited only to the periphery. The appearance was consistent with a cardiomyopathy called " Arrhythmogenic Right Ventricular Dysplasia (ARVD)" (**Fig 1**)

No evidence of aspiration, pulmonary thromboembolisation and pulmonary hypertension were noted in lung tissues.

The histology of other organs was unremarkable.

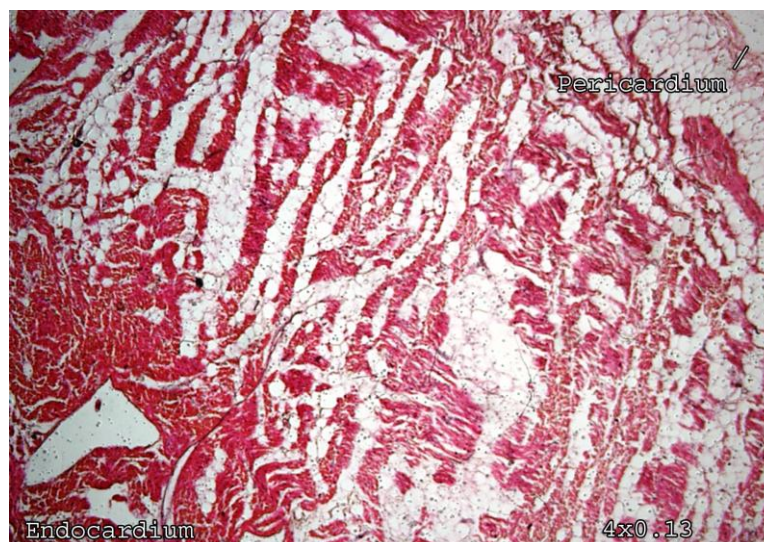


Figure 1: Microphotograph of the full thickness of right ventricular myocardium neck magnification

Discussion

Sudden deaths caused by both Arrhythmogenic Right Ventricular Dysplasia (ARVD) and persistent Atrial Septal Defects (ASD) have been reported in the medical literature.^{1,2} In the absence of other demonstrable pathology and after reviewing the circumstances leading to the death, it is prudent to come to the conclusion that this death can be attributed to ARVD. It can be considered as a Class II Cause of Death where there is a potentially fatal pathology with suggestive circumstances.³ It can also be considered as a decision which can withstand to the criticism of those looking for an anatomic cause of death as well as the standard required in legal procedure if necessary.^{3,4}

ARVD causes arrhythmias in children and young adults. Only 30-50% has a familial distribution.² It is mainly inherited in an autosomal dominant manner, with variable expression. The penetrance is 20-35% in general.⁵ Its clinical presentation has four stages and initial asymptomatic stage is called the concealed phase.²

The concealed phase is characterized by subtle right ventricular changes with or without minor ventricular arrhythmias. Detection of ARVD especially in this phase is difficult even with sophisticated investigations such as myocardial biopsy.⁵ Unfortunately the first manifestation of this phase may be sudden death.^{5,6,7}

Other advanced phases are usually having signs and symptoms.^{5,6} Undiagnosed ARVD has been reported to have caused sudden post operative deaths.¹

Histologically ARVD has two distinct patterns, namely fatty infiltration and fibro-fatty infiltration.^{5,6}

Fatty infiltration is first confined to the right ventricle. There is partial or near-complete substitution of myocardium with fatty tissue without obvious thinning of the myocardium. It involves predominantly the apical and infundibular regions of the right ventricle. The left ventricle and septum are usually spared. But in later stages it can extend to the left ventricle too. No inflammatory infiltrate is seen.^{5,6} This is the histological variety found in this case.

In fibro-fatty infiltration, myocytes are replaced with fibrofatty tissue with inflammatory infiltrate. There can be thinning of the right ventricular free wall (< 3 mm thickness).

90% of individuals with ARVD have ECG abnormalities and the most common ECG abnormality is non specific T inversion in leads V₁ to V₃. The epsilon wave (a terminal notch in the QRS complex of ECG) is found only in 50%.⁵

Echocardiogram may show hypokinesia in affected walls and Magnetic Resonance Imaging (MRI) may show changes in density in myocardium.⁷

Biopsy will give histological diagnosis provided it has taken exactly from affected site.^{5,6,7} It should also be appreciated that ARVD has very variable presentation.⁶

Atrial septal defect is also known to cause atrial arrhythmias especially during hypoxic conditions. Usually ASD >2cm causes reversal of shunt and

pulmonary hypertension. ASDs can be free of murmurs and may be detected only in very late life.²

Usually only ECG, Chest X ray and relevant blood tests are done preoperatively on cardiologically asymptomatic young patients in government hospitals in this country. As far as surgery, anaesthesia and postoperative management are concerned, it seems that the clinicians have followed the accepted practice of our public health care system. Therefore success of any medical negligence allegation can be considered very remote.⁴

References

1. Khoy-Wee Toh et al, Postoperative death in a patient with unrecognized ARVD syndrome, *Anaesthesia and analgesia*, 2004; 99: 350-2.
2. Babara A.M. et al. An unusual presentation of ASD in a patient undergoing total hip replacement. *Anaesthesia and Analgesia*, 2000; 91:1134-1136.
3. Adems V.I, Flomenbaum MA, Hirsch CS, Truama and Disease. In: Spitz DJ, Spitz WU eds. *Spitz and Fisher's Medico-legal Investigation of Death*, Illinois: Charles C Thomas Publishers Ltd, 2006: 455-49
4. Fernando R, De Silva L.C. *Basic Principles of Medical Negligence*. Colombo: The Institute of Health Studies, 1996
5. Arrhythmogenic right ventricular dysplasia. http://en.wikipedia.org/wiki/Arrhythmogenic_right_ventricular_dysplasia. (retrieved on 07/11/2011)
6. Arrhythmogenic right ventricular. Dysplasia. Gear K, Marcus F, *Circulation*. **2003**; 107: **e31-e33**(retrieved 07/11/2011)
7. Imaging in Arrhythmogenic Right Ventricular Dysplasia. Lawrence A, Eugene C. L www.emedicine.com/radio/topic903.htm (retrieved 23/04/2011)