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Arrhythmogenic Right Ventricular Dysplasia: How to find what you are not looking for?

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Abstract

Arrhythmogenic right ventricular dysplasia is a life-threatening condition. It can be asymptomatic during the first decades of life and debut as ventricular arrhythmias or sudden death. We report a 32-year-old man, scheduled to perform a surgery due to a fracture of the left radius, who was diagnosed with the disease of arrhythmogenic right ventricular dysplasia. With the present case, the aim is to publicize the characteristics of the disease and its anesthetic management in order to establish an early diagnosis and to be able to take adequate precautions in the operating room.

Case study

We report a 32-year-old man, scheduled to perform an osteosynthesis due to a fracture of the left radius with a previous diagnosis of Arrhythmogenic Right Ventricular Dysplasia (ARVD). He was a high competition athlete until he was 19 years old and had an uncertain family history of ARVD. He did not take any pharmacological drugs and did not carry a pacemaker. He was diagnosed by Magnetic Resonance Imaging (MRI) (Figure 1) finding of right ventricular dilatation because he felt palpitations accompanied normal activity. Echocardiography showed a Left Ventricular Ejection Fraction (LVEF) of 43% and systolic dysfunction mild, dilated right cavities and moderate tricuspid insufficiency with PSAP of 25-30. In the theatre standard monitoring was used and it was available an external defibrillator. An ultrasound-guided block of the axillary plexus was performed with levobupivacaine and mepivacaine. Some drugs as amiodarone and beta blockers were initially prepared but the patient remained hemodynamically stable during perioperative period and any arrhythmia was detected.

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ARVD is an inherit cardiomyopathy in which the cardiomyocytes are replaced by fibro-fatty tissue, mainly in the right ventricle. It causes cardiac failure and life-threatening ventricular arrhythmias. It is an autosomal dominant pathology with variable penetrance, which occurs in about *1 in 5000 people*, so it is common for an anaesthesiologist to find a patient with this disease throughout his professional career [1]. Young men are 3 times more affected than women. The symptomatology begins in adolescence, and it is diagnosed around the age of 30. The disease manifests as palpitations, dyspnoea, fainting, ventricular arrhythmias or Sudden Cardiac Death (SDC), usually during physical efforts.

There is currently no specific diagnostic test. It is taken into account some major criteria such as family history of the disease, evidence of electrocardiographic abnormalities (T wave inversion in V1-V3, right branch block, epsilon waves, ventricular tachycardia…), the presence of cardiac dysfunction in imaging tests and fibro-fatty replacement in RV. Cardiac magnetic resonance and 3D echocardiography are preferred to right ventricular angiography for assessing morphology and functional abnormalities of the right ventricle [1-3]. Regarding the treatment, drug therapy is attempted in symptomatic patients or to prevent arrhythmias. An automatic defibrillator (ICD) is recommended in patients with severe clinical or ventricular tachycardia.

ARVD causes a high rate of unexplained perioperative deaths in anaesthetised patients. It constitutes one of the main causes of sudden cardiac death in apparently healthy young adults [1,2]. The under diagnosis is the mayor vulnerability of this pathology even more in the operating room. Furthermore, there is a small number of published literature about its anaesthetic management. Before the intervention it is recommended to have recent ECG and echocardiogram and not to stop the antiarrhythmic medication if patients are taking it. There is no contraindication for performing general or regional anaesthesia in this type of patient. Nevertheless, the use of adrenaline and high doses of bupivacaine in regional techniques should be avoided [3].

During the surgical intervention the best strategy to prevent the onset of ventricular arrhythmias is the maintenance of proper antiarrhythmic treatment and optimal hemodynamic conditions for myocardial perfusion. The provision in the operating room of amiodarone and a defibrillator device is recommended in case intraoperative arrhythmias occur. In the postoperative period, more intensive and continuous surveillance is recommended.

As noted above, the big problem of these rare diseases is the under diagnosis and the poor knowledge of these pathologies. Therefore, more research should be done in order to avoid deaths due to these reasons. Currently, Apps are being developed whose objective is to make a strategy to, quickly and effectively, look up for the perioperative risks of patients with rare diseases and share ever experience in order to design an algorism [4]. Nevertheless international data sharing initiatives in health and research for rare diseases need also appropriate legislations in order to respect patients' privacy and needs for information regarding the use of their data [5].

Conclusion

To conclude, note that the ARVD is a not infrequent entity, which can cause high morbidity and mortality in anaesthetised young patients. A careful perioperative evaluation and the establishment of a management plan focused on the conservation of physiological stability helps to avoid complications and provide safety in this population. It is important to share our experience but also avoid publishing patient privacy data.

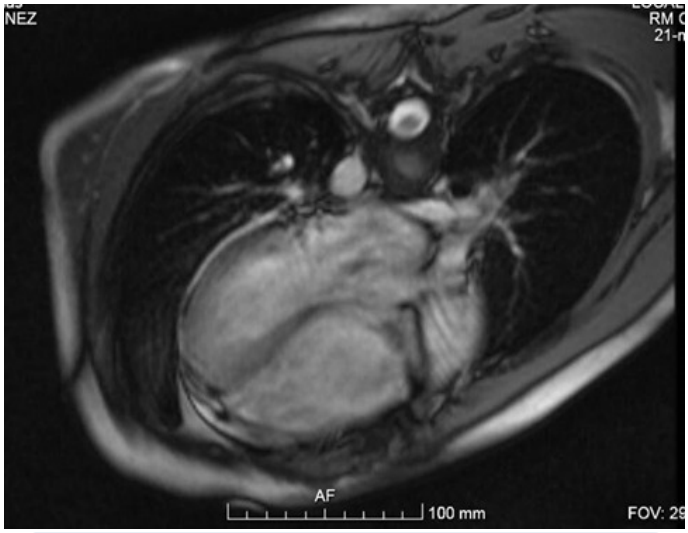

Figure 1: Cardiac Magnetic resonance imaging. Dilated right cavities.

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