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Anomalous left coronary artery from the pulmonary artery (ALCAPA) as a concealed cause of severe mitral regurgitation in children and adults: Case reports

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ABSTRACT

INTRODUCTION: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital coronary artery anomaly and is a concealed cause of mitral regurgitation.

PRESENTATION OF CASE: We reported two cases of severe mitral regurgitation in whom the presence of ALCAPA was overlooked in the first cardiac surgery. In the first case, ALCAPA was diagnosed one year after the mitral annuloplasty and the surgical reimplantation was successfully performed. In the second case, ALCAPA was incidentally detected on multislice computerized tomography during preoperative workup for the second surgery to replace the dehiscent mitral prosthesis.

DISCUSSION: The clinical presentation of ALCAPA varies highly. It is not difficult to diagnose an ALCAPA in newborns and infants. Contrarily, ALCAPA in children and adults can be overlooked.

CONCLUSION: Physicians should always look for the presence of ALCAPA in patients who present with unexplained mitral regurgitation. The surgical strategy in patients with ALCAPA should be carefully individualised to achieve an optimal outcome and alleviate complications.

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1. Introduction

Anomalous left coronary artery from the pulmonary artery (ALCAPA) was first described by Brooks in 1885 [1] and possibly in the 1860s by Krause [2]. ALCAPA is a rare congenital anomaly of the coronary artery [3–5]. There are two types of ALCAPA: the infantile type and the adult type, each of which is characterised by different pathophysiology, clinical manifestations, surgical treatment, and outcomes [6]. ALCAPA can be the cause of dyspnea, chest pain, dysrhythmia, or even sudden death in adults [2,6–8]. ALCAPA has been sporadically reported as a rare cause of mitral regurgitation (MR) [9].

In this paper, we report two patients with severe MR. Both underwent previous cardiac surgeries to deal with the MR. Unfortunately, the ALCAPA was not recognised during the first operations. Our two cases emphasised the crucial role of the clinical vigilance in looking for a hidden ALCAPA in patients with MR. The strategy for corrective surgical repair in adult patients with ALCAPA

should be individualised to achieve an optimal outcome and avoid complications.

The article has been reported in line with the SCARE criteria [10].

2. Presentation of case

2.1. Case 1

A 2-year-old girl was referred to our centre for dyspnea on exertion and failure to thrive. Echocardiography revealed a left ventricular (LV) ejection fraction of 56% and a severe MR attributed to congenital shortened chordae tendinae and mitral annular dilatation. She underwent a cardiac surgery for mobilisation of chordae and an autologous pericardial strip mitral annuloplasty. Following the operation, the MR was reduced and rated as mild. The patient was discharged without any issues. The heart failure symptoms became less pronouncing but her physical function did not reach the expected level. The echocardiographic checkup performed one year after her operation suggested an ALCAPA: a dilated right coronary artery (RCA), reversal flow in the left main coronary artery (LMCA) (Fig. 1A), an abnormal flow from LMCA to the main pulmonary artery (Fig. 1B), and an increased right-to-left coronary collaterals (**Supplement video 1**).

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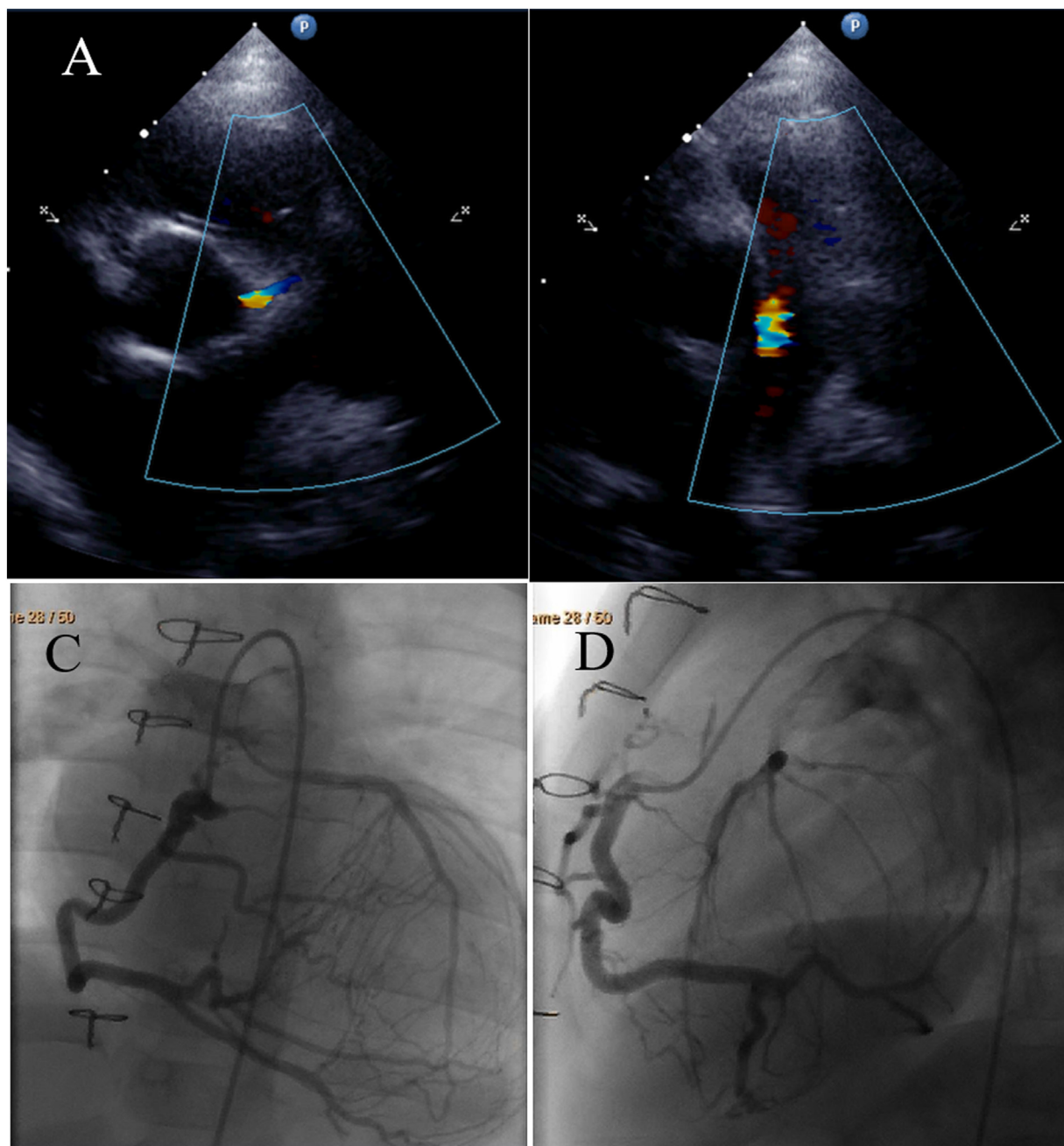


Fig. 1. Echocardiographic and angiographic findings of the first case. A reversed flow in the LCA (A) and an abnormal flow to the MPA (B) were seen. The angiogram (C and D) showed a dilated RCA, diffuse collaterals, and the backflow in the LMCA.

The coronary angiography confirmed the ALCAPA. The coronary arterial collaterals drained blood from the dilated RCA to the left circumflex (LCx) and the left anterior descending artery (LAD) and back to the MPA (Fig. 1C-D, Supplement video 2, and Supplement video 3). The surgical reimplantation of the LMCA to the aorta was performed with success. The patient was discharged seven days following the operation. The most recent echocardiography showed a good LV ejection fraction of 58%, a normal LV global longitudinal strain, a trace MR with minor hyperechogenic spots on the chordae tendineae (Fig. 2).

2.2. Case 2

A 32-year-old female, mother of two children, was referred to our centre after an exertional fainting. She underwent two urgent

Cesarean sections because of a severe rheumatic MR leading to the imminent acute pulmonary oedema during her labors. She subsequently experienced a mitral valve replacement. She reported a functional improvement for the first year after the operation, but then her health status declined gradually. She was on high doses of antidiuretics, angiotensin-converting enzyme inhibitor, digoxin, and a beta-blocker.

Physical examination revealed: body weight 50 kg, height 150 cm, BMI 22 kg/m², heart rate of 60–80 beats/min, blood pressure 115/60 mmHg, respiratory rate 18/min. She had 3/6 degrees systolic murmur at the cardiac apex. The electrocardiogram confirmed an atrial fibrillation. The echocardiography showed a dilated LV (LV end-diastolic diameter 64 mm), poor LV contraction (LVEF 40%), and a mitral mechanic prosthesis with profuse dehiscence

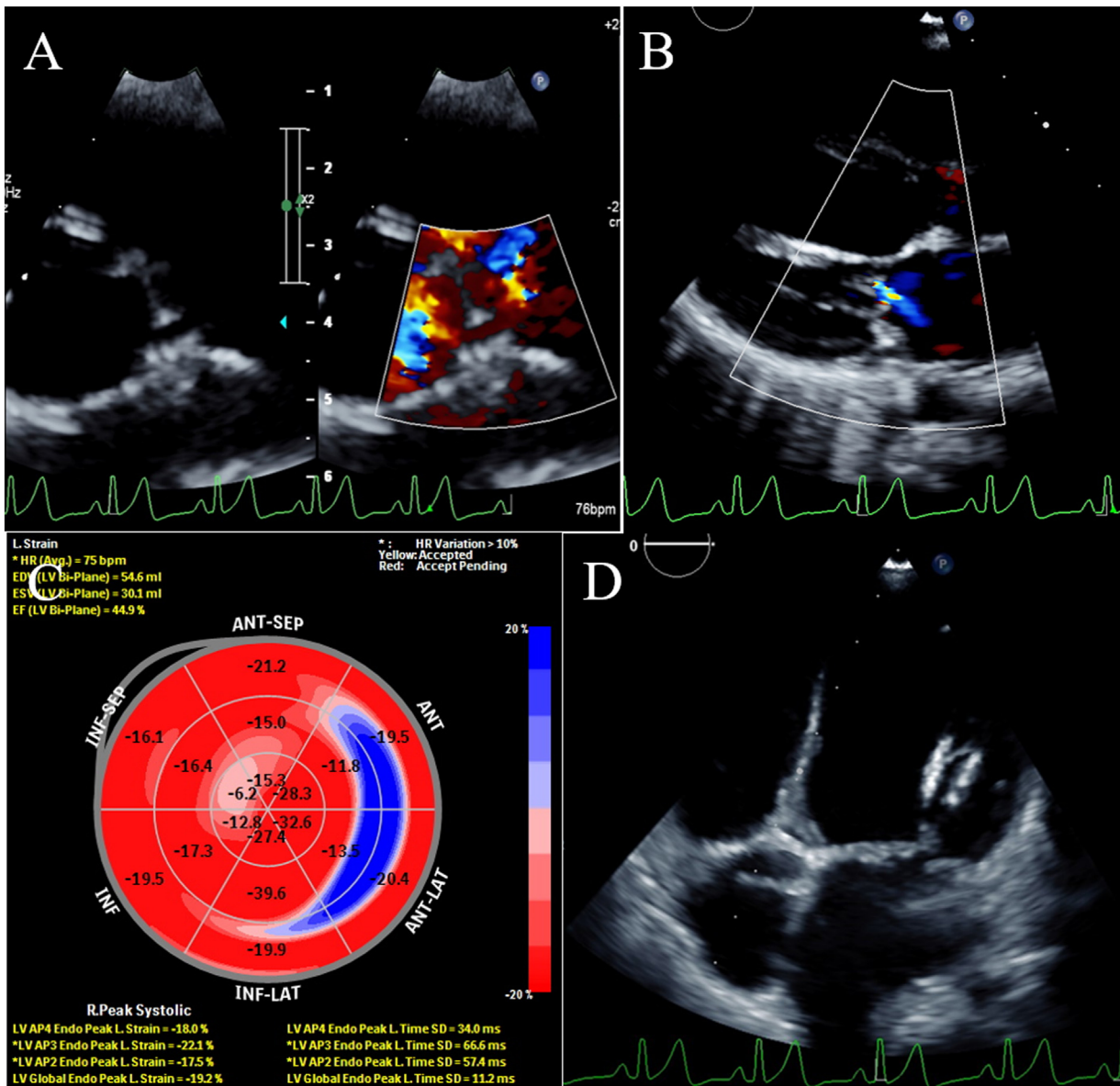


Fig. 2. Echocardiographic findings following the reimplantation of ALCAPA in the first case. The normal flow in the LMCA was reestablished (A). A mild MR (B) and a normal LV strain with mild reduction at the LMCA territory (C) were noted. Hyperchogenic chordae tendinae were seen (D).

causing a severe paravalvular leak (Fig. 3A), and rocking movement (Supplement video 4).

A second cardiac surgery was planned to replace the dehiscant mitral prosthesis. Due to the previous surgery, a thoracic multislice computer tomography was performed. To our surprise, an ALCAPA was detected: a severely dilated RCA, an anomalous LMCA from the main pulmonary artery, and increased coronary collaterals (Fig. 3B–D). The surgical plan was to replace the dehiscant mitral prosthesis, ligate the proximal LMCA, and coronary artery bypass grafting (CABG). Bicaval cannulation was conducted to achieve adequate venous return and exposure of the pulmonary artery. Retrograde cardioplegia with Custodiol solution was delivered via the coronary sinus. Recognizing the significant calibre mismatch between the small internal mammary artery and the dilated LMCA, the surgeon decided to reimplant the anomalous LMCA instead of CABG. The first attempts of weaning from cardiopulmonary bypass were difficult with reiterative ventricular fibrillations requiring repeated epicardial shocks. In addition, the bleeding from the LMCA anastomosis to aorta needed repeated explorations. The patient was not

able to be weaned from cardiopulmonary bypass. She was declared dead after 9 h of cardiopulmonary support.

3. Discussion

The diagnosis of ALCAPA in neonates is straightforward. However, the recognition of atypical ALCAPA in children and adults is more challenging. In our two cases, the real etiology of MR was missed during the first surgery. For the first case, a severe MR and a good LV contraction before the first operation might mislead the heart team. The LV function is dependent on the degree of interarterial coronary collaterals [4,6], which developed well in this patient. The hyperchogenic chordae tendinae of the mitral valve did trigger a further investigation aiming at ALCAPA. The second case was presented with predominant signs and symptoms of heart failure and mitral prosthetic dehiscence without any written document from previous surgery. In this patient, we were not aware of the presence of an ALCAPA, which seemed to be the cause of MR leading to the previous operation. The ALCAPA was incidentally diagnosed. Dahle and coworkers recommended that ALCAPA must be excluded

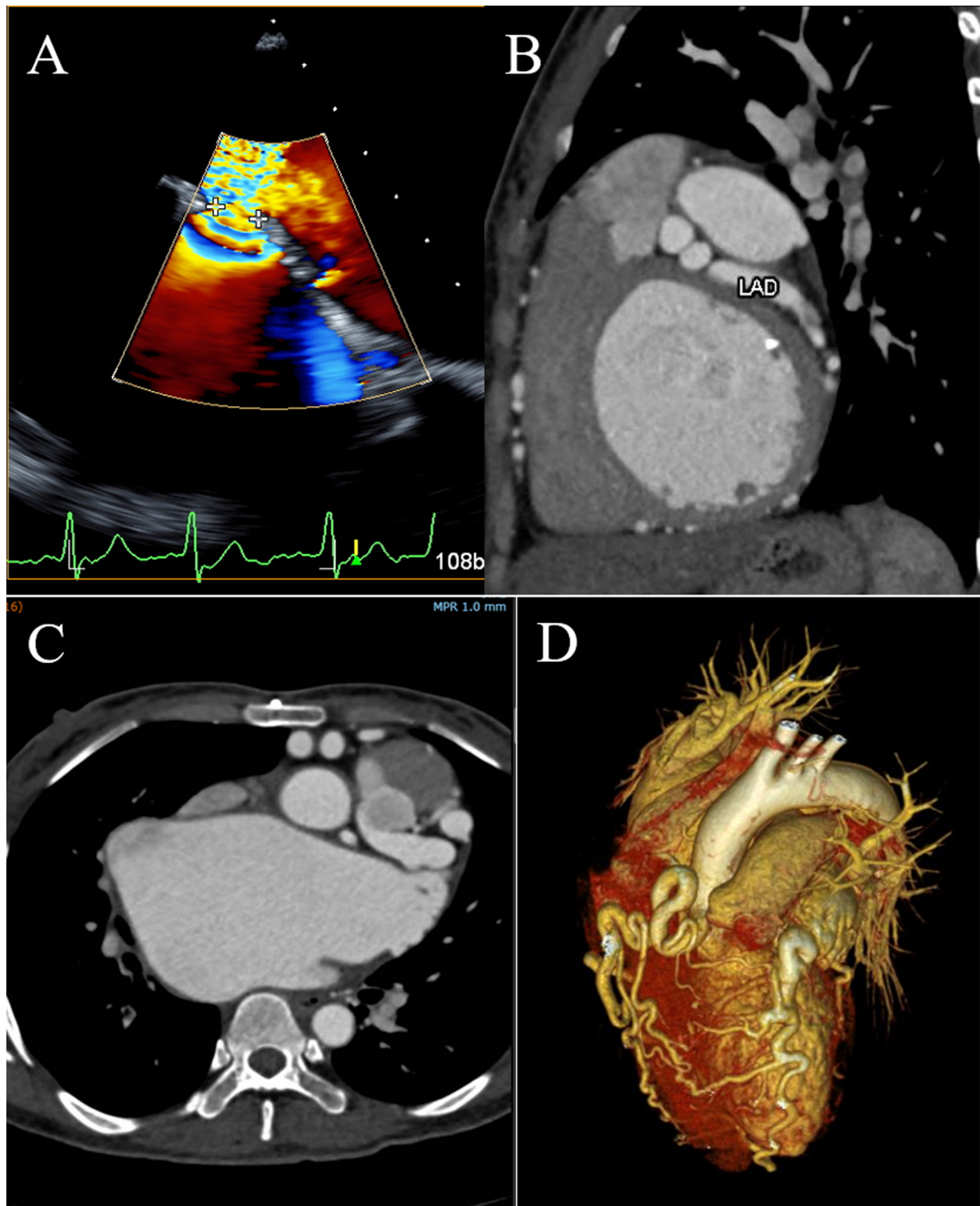


Fig. 3. Echocardiographic and angiographic findings of the second case. A severe paravalvular leak was seen (A). The LAD trajectory (B) and the contrast from LAD entering the main pulmonary artery (C) showed on MSCT. The 3D reconstruction showed a dilated RCA, and diffuse collaterals (D).

in younger patients with MR and no other morphological findings [9].

While early direct reimplantation of the anomalous LMCA into the aorta is the treatment of choice for infantile type [11], the optimal surgical management of ALCAPA in adults is still a debatable issue [2]. Various surgical methods have been introduced, including ligation of the anomalous LMCA, CABG, Takeuchi's procedure, and reimplantation of the LCMA to aorta [12]. Wilson and coworkers

compared two surgical approaches: simple ligation versus ligation in combination with CABG, and found no significant difference in survival between the two groups [13]. Given the fact that ALCAPA in adults is a rare condition and the technique for implantation of LCMA was challenging, we had planned to replace the dehiscent mitral prosthesis, ligate the anomalous LMCA, and perform the CABG. However, in fear of calibre mismatch and competitive flow, the surgeons changed their mind and performed the reim-

plantation. This strategy might lead to bleeding and prolong the ischemic time in a heart already weakened by a longstanding disease. In this case, the ligation of the anomalous LMCA at its origin in combination with CABG would be more appropriate.

4. Conclusion

ALCAPA is a rare congenital anomaly of the coronary artery. The manifestation of ALCAPA in older children and adults can be atypical and misleading. Although rarely seen, ALCAPA is a proven cause of severe MR. In patients with severe MR, the clinicians should be aware of all possible etiologies of MR, including ALCAPA. Surgical strategy for ALCAPA in adults must be carefully individualized.

Declaration of Competing Interest

The authors declare that there is no conflict of interest.

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Ethical approval

The manuscript was approved by the Institutional Review Board of the University Medical Center Ho Chi Minh City, number 167/BVDHYD-HDDD, on July 16th 2020.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Dr. Nguyen Hoang Bac: study concept, data collection, drafting the first manuscript.

Dr. Cao Dang Khang: data collection, contribution in writing the first manuscript.

Dr. Pham Thi Hieu Thao: data collection, data analysis.

Dr. Vo Tuan Anh: data collection, data analysis.

Dr. Le Minh Khoi: study concept, data collection, data analysis or interpretation, writing the final manuscript.

Registration of research studies

N/A.

Guarantor

Dr. Le Minh Khoi, M.D.

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Appendix A. Supplementary data

Supplementary material related to this article can be found, in the online version, at <https://doi.org/10.1016/j.ijscr.2020.09.022>.

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