Case report

Abnormal drainage of inferior vena cava to left atrium combined with atrial septal defect: A case report

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ABSTRACT

Systemic venous anomaly is less common clinically, and ectopic connection of inferior vena cava is less common than ectopic connection of superior vena cava. When the inferior vena cava is abnormally connected to the left atrium, a low atrial septal defect can be combined. It is difficult to make preoperative diagnosis of this kind of disease clinically. Blind occlusion of the ASD (atrial septal defect) will cause the iatrogenic inferior vena cava flowing back to the left atrium completely or partially. Conventional median thoracotomy for repair of such an atrial septal defect should be performed with more caution. It has been reported that after repair of an atrial septal defect, the inferior vena cava was mistakenly separated into the left atrium. Therefore, we suggest that preoperative evaluation of inferior atrial septal defect should be with more cautious. When transthoracic echocardiography (TTE) cannot fully explain the clinical symptoms, Ultrasound contrast may be considered. During the repair of the inferior vena cava ASD, it is not only necessary to find the location of Euclidean valve, but also to accurately explore the inferior vena cava opening so as to avoid the iatrogenic inferior vena cava being separated into the left atrium.

1. Introduction

The technique of interventional closure of atrial septal defect is relatively mature, but there have been reports of iatrogenic right-to-left shunt after interventional closure, which may be related to the malformation in this report. Therefore, the abnormal connection of inferior vena cava should be carefully determined during interventional plugging. Preoperative echocardiography and angiography were used for accurate diagnosis if necessary.

2. Case report

The patient was a 22-year-old male with a height of 185 cm and weight of 100 kg. An atrial septal defect (ASD) was found by cardiac ultrasound during a health examination 10 months ago. Transthoracic echocardiography (TTE) showed that there was an echo interruption in the posterior lower part of the atrial septum near the entrance of the inferior lumen, the size of which was about 2.0 cm. The distance from the anterior lower edge of the defect to the root of the anterior mitral valve was about 3.68 cm, and to the entrance of the superior vena cava was about 1.73 cm. There was no obvious stump at the posterior lower part of the atrial septum and the entrance of the inferior vena cava. CDFI: Left to right shunt signal was detected at the atrial septal defect, and blood flow from the right inferior pulmonary vein entered the left atrium and entered the right atrium homeopathically through the atrial septal defect. Considerations: congenital heart disease, atrial septal defect (inferior cavity type) (Fig. 1). Six months ago, the reexamination of cardiac ultrasound was considered as congenital heart disease, atrial septal defect (ostium secundum defect), the defect was about 34 mm, 35 mm, 21 mm and 5 mm from the anterior mitral valve, aortic valve and the entrance of superior and inferior vena cava respectively, and there was no stump at the apex of the atrial septum. The atrial level was shunted from left to right. The patient's daily activities were not affected, and he denied the history of chest tightness, dyslexia, cyanosis, and repeated pneumonia. Physical examination: no obvious cyanosis, no clubbing fingers. The heart apical fluctuation was located about 2 cm above...
outside the 5th intercostal area of the left midline of the clavicle, the heart rate was 80 beats/min, sinus rhythm. Soft systolic blowing murmur could be heard in the 2nd and 3rd intercostal areas of the left edge of the sternum, while no obvious pathological murmur in the other valve areas. Preoperative blood gas analysis (36.5 °C, without oxygen intake) suggested PaCO\(_2\) 35.3 mmHg, PaO\(_2\) 64.6 mmHg, Lac 1.9 mmol/L, SaO\(_2\) 93.0 %. The electrocardiogram suggested sinus rhythm. Interventional occlusion was tried firstly, the guide wire and angiography catheter were placed and successfully entered the left superior pulmonary vein (Fig. 2). The angiography showed a left to right shunt with rapid development of the right atrium at the same time. After attempting to release the occluder, the shape was abnormal and could not be fixed on the atrial septum, then the septal defect closure was abandoned. Patch repair of atrial septal defect through right thoracic small incision assisted by cardiopulmonary bypass was performed. Extracorporeal circulation was established through femoral arteriovenous intubation. Right internal jugular vein intubation was used for auxiliary drainage. Circulation was conducted in parallel, and small incision was made into the chest through the right anterolateral incision, and carbon dioxide continued to be injected into the operative field. The inferior vena cava was obviously widened and it was difficult to block. After blocking the superior vena cava, the right atrium was directly incised, and the atrial defect was found to be inferior vena cava type, about 20 mm. The stump of the lower edge is about 5 mm. The inferior vena cava intubation was located in the left atrium, and it is adjacent to the right inferior pulmonary vein. The pulmonary veins were all opened in the left atrium, and the coronary sinus was opened in the right atrium. The superior vena cava developed and connected normally, and the left superior vena cava was not found. The tricuspid valve was normal and the annulus was not enlarged. Abnormal inferior vena cava connection into the left atrium with an atrial septal defect was considered. The lower edge of the atrial septal defect was cut off, and the atrial septal defect was enlarged and cut upward to the fossa ovale. The posterior wall of the left atrium was sutured with a suitable size polyester patch. The inferior vena cava was separated into the right atrium, and the opening of the right inferior pulmonary vein was protected. The atrial septal defect was closed and the inferior vena cava orifice could easily pass through the 22# right ventricular outflow tract probe. ECG indicated sinus rhythm. Postoperative ultrasound revealed no shunt at the level of atrial septum and no obvious dilation of inferior vena cava. He was discharged 7 days after surgery. Six months and one year after surgery, cardiac ultrasound was shown in Figs. 3 and 4. There was no shunt at the level of atrial septum and no obvious dilation of inferior vena cava.

3. Discussion

Abnormal connection of the body vein is less common in clinical practice. Abnormal connection of the inferior vena cava is rarely than that of the superior vena cava. The incidence of abnormal connection of the inferior vena cava is about 0.6 % in congenital heart disease. Abnormal connection of the inferior vena cava includes absence of the inferior vena cava and abnormal connection of the inferior vena cava into the left atrium. However, when the inferior vena cava is abnormally connected to the left atrium, low atrial septal defect can be combined. Such children are often accompanied by ectopic drainage of the right pulmonary vein or common atrium [1]. The patient underwent a small right thoracic incision surgery, through the right femoral vein intubation, the inferior vena cava was found to be very wide and deep intraoperatively, which was difficult to be bridled and blocked. Therefore, instead of forcibly blocking the inferior vena cava, the right atrium was

![Fig. 1. Congenital heart disease, atrial septal defect (inferior cavity type).](image-url)
directly cut open after blocking the superior vena cava. The femoral vein intubation orifice is located in the left atrium visibly. The diagnosis of atrial septal defect was clear. The lower margin of the defect is located right anterior to the opening of the inferior vena cava. The lower margin of the atrial septal defect was sufficiently enlarged intraoperatively, and the Dacron patch was sutured with the posterior wall of the left atrium to segregate the lower cavity back to the right atrium and repair the atrial deficiency.

Such patients with accurate preoperative diagnosis are difficult. Transthoracic echocardiography (TTE) is easy to miss diagnosis. Inferior vena cava angiography has a high accuracy, with left cardiac system imaging rapidly, but it still needs to be differentiated from bidirectional or right-left shunt inferior atrial septal defect. Ultrasound indicated left-to-right shunt in this patient. It was found that the inferior vena cava flowed back into the left atrium and quickly entered the right atrium through the defect intraoperative, and the left heart system was not image developed, which limited the accurate diagnosis of interventional sealing. In addition, cyanosis is not obvious and the low PaO$_2$ in pre-operative blood gas analysis could not fully explain by the atrial septal defect of the left to right shunt, which should be paid attention seriously. Contrast-enhanced ultrasound may be considered for further clarification [2]. In terms of treatment, for inferior atrial septal defect, interventional occlusion must be performed carefully. Firstly, the right inferior pulmonary vein should be paid more attention to prevent obstruction and then more attention should be paid to the rare abnormal connection of inferior vena cava, so as to prevent the iatrogenic inferior vena cava from completely or partially returning to the left atrium after blind occlusion. Routine thoracotomy for this type of atrial septal defect should be more cautious. There have been reports that inferior vena cava was accidentally isolated into the left atrium after patch repair of atrial septal defect [3]. During the median of thoracotomy, we considered that the superior vena cava pouch and occlusion were normal, but the inferior vena cava pouch was often sutured in the epicardium at the junction of the inferior vena cava and the right atrium, and it was difficult to detect the opening of the inferior vena cava in the heart after intubation occlusion. Abnormal connection of inferior vena cava to the left atrium was missed. Mistaken the Euclid valve for the lower margin of the ASD. Actually, inferior vena cava opening was located in the left atrium, and after the removal of the inferior vena cava cannula the inferior vena cava opening was still located in the left atrium while tightening the purse of

![Fig. 2. The catheter was successfully inserted into the left superior pulmonary vein during interventional closure.](image1)

![Fig. 3. Six months postoperative follow-up.](image2)
the epicardial membrane, which resulting in iatrogenic right-to-left shunt. In this case, if the patient underwent median thoracotomy, it was necessary to carefully explore the intracardiac malformation, otherwise, the above errors would easily occur. Probing the inferior vena cava opening is not affected by intubation through the femoral vein.

4. Conclusion

Preoperative evaluation of inferior atrial septal defect should be cautious. Ultrasound contrast or venous angiography should be considered when transthoracic echocardiography (TTE) cannot fully explain the clinical symptoms. Intraoperative repair of the inferior vena cava deficiency, we should not only look for the Eustachian valve position, but also explore the opening of the inferior vena cava accurately, so as to avoid iatrogenic inferior vena cava separation into the left atrium.

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Ethical approval

The patient in this case report is our regular patient. For these patients, we signed the “Scientific Research Informed Notification notice” and the patients agreed to use their medical information for scientific research and publication. The medical ethics Management Committee of Peking University Medical Lu Hospital of Traditional Chinese Medicine approved the data for scientific research.

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

Zheshu Li, Ph.D, performed the procedure and reviewed the whole process of paper publication.

Xingming Wang involved in interpretation and collecting of data, writing- Original draft of the manuscript and revising it.

Mei Zhou, Lei Wang and Lei Han involved in writing Original draft of the manuscript. Peng Li involved in editing the final version of manuscript.

All authors reviewed the paper and approved the final version of the manuscript.

Registration of research studies

Not applicable.

Guarantor

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Declaration of competing interest

There is no competing of interest to report.

References

[1] Zhu Xiaodong, Zhang Baoren (Eds.), Cardiac Surgery, People’s Medical Publishing House, Beijing, 2007, pp. 354–366.
[2] Junmin Quan, Qianmin Tao, Wu. Bifeng, et al., Diagnosis of abnormal inferior vena cava-left atrial tunnel after operation of atrial septum defect accompanied with partial anomalous pulmonary venous connection by contrast-enhanced ultrasound:a case report(J), J. Ultrasound Clin. Med. 18 (02) (2016) 123–129.

[3] Xu. Zhong-Ying, Wei-Li Yang, Yan Zhang, et al., Imaging diagnosis of inferior vena cava drainage into the left atrium after atrial septal defect (report of 3 cases), J. Clin. Radiol. 07 (2001) 558–559.

[4] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, International Journal of Surgery 84 (2020) 226–230.