Cardiology in the Young

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Surgical treatment of cardiac tumour thrombus in children

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Brief Report

Cite this article: Yi H, Li X, and Wang H (2023) Surgical treatment of cardiac tumour thrombus in children. *Cardiology in the Young* **33**: 2443–2445. doi: 10.1017/S1047951123002652

Received: 29 May 2023
Revised: 27 June 2023
Accepted: 27 June 2023
First published online: 25 July 2023

Keywords:
Cardiac tumour thrombus; surgical treatment; children

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Abstract

Cardiac mass in children is rare and insidious onset, and primary cardiac mass is less than secondary mass. Among the malignant tumours in children with tumour thrombus in the venous system, about 98% of the cases are nephroblastoma. But it is still rare for the tumour thrombus to reach the level of the atrium or even enter the right ventricle. In this case, the child complained of chest tightness and palpitation and went to the doctor and found Wilms tumour complicated with intracardiac tumour thrombus.

Cardiac mass in children is rare, and the main types are benign tumours, malignant tumours, tumours with unknown biological behaviour, or non-neoplastic lesions.[1] Cardiac metastases have an incidence of 0.7–3.5% in the general population (up to 9.1–18.3% in patients with known cancer) and are more frequent than primary cardiac tumours (incidence of 0.01–0.3%).[2,3] The clinical manifestations of cardiac mass are often atypical, and many of them are discovered incidentally during the examination of other diseases. The appearance of relevant symptoms may indicate that the mass has caused haemodynamic changes. In this case, the child complained of chest tightness and palpitation and went to the doctor and found Wilms tumour complicated with intracardiac tumour thrombus.

Case presentation

The patient was a 4-year-old boy with insidious onset. He visited the doctor in local hospital because of palpitation, chest tightness, shortness of breath, and abdominal distension, and physical examination revealed a palpable right abdominal mass. So, echocardiography, abdominal ultrasonography, and urinary ultrasonography were taken and revealed mass-like hyperechoic echo can be seen in the right atrium, which starts from the inferior vena cava and fills the entire right side, with a size of about 47*39 mm. The inner diameter of the entire inferior vena cava was widened, and the cavity was filled with a strong echogenic mass, the widest part was 25 mm. The volume of the right kidney was enlarged, and a heterogeneous cystic mass with a size of about 96*71 mm and a poorly defined border was very visible in essence. And then an enhanced abdominal CT was taken. That revealed a huge cystic and solid mass was seen in the right kidney area, with a size of about 7.8*7.5*5.4 cm. The boundary between the mass and the enlarged right renal parenchyma was unclear, the right renal pelvis and calices were compressed, and the structure of the right adrenal gland was not clearly displayed, the inferior vena cava is widened, and its internal density is uneven. Strips and slightly low-density shadows can be seen in the right atrium. The enhancement shows that the volume of the right kidney is increased, and an obvious uneven enhancement mass shadow can be seen. The mass is embracing the adjacent remnant kidney spherical. Filling defects can be seen in the right renal vein, inferior vena cava, and right atrium, with different degrees of enhancement. According to these results, local doctors first considered that's primary renal tumour with cardiac tumour thrombus may probably be Wilms tumour. A needle biopsy of the tumour was performed on the child and diagnosed Wilms tumour (mixed type).

After diagnosis, chemotherapy was given as soon as possible. Alternating chemotherapy with "vincristine, actinomycin" and "vincristine" was given for 4 weeks and then ultrasound evaluated the size of the tumour thrombus, but it was still located in the atrium. So we added chemotherapy "vincristine, actinomycin, doxorubicin" twice, once every 3 weeks. Then ultrasound evaluated the right nephroblastoma has shrunken, but the right renal vein and inferior vena cava tumour thrombus (full type) extended to the right atrium and protruded to the right ventricle. Pre-operative echocardiography showed that the intracardiac tumour thrombus was located in the right atrium, travelling to and from the right atrium and right ventricle with the heartbeat (crossing the tricuspid valve during diastole), which slightly affected the blood flow of the tricuspid valve. An enhanced chest and abdominal CT were also showed that (Fig. 1). So at that time, surgery was the only option. So he was transferred to our hospital.

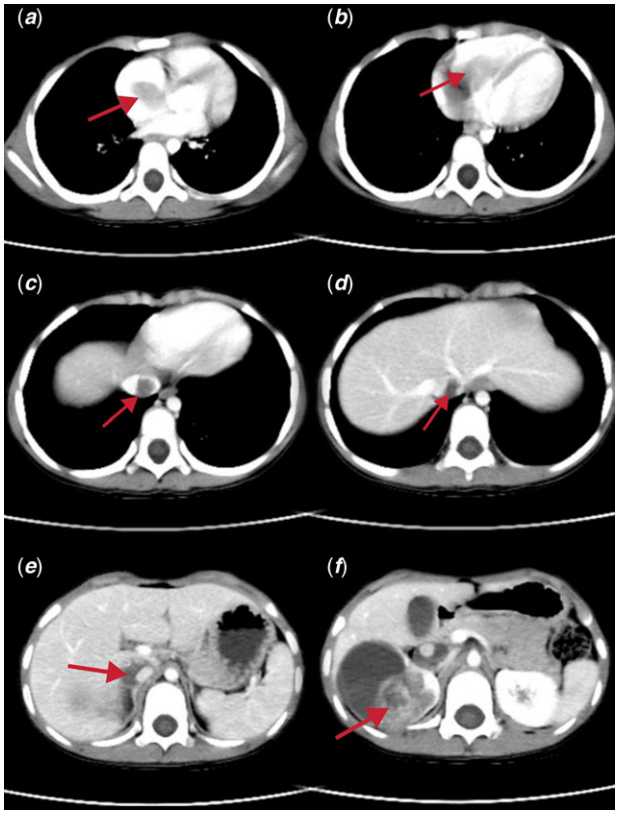

Figure 1. Chest and abdomen CT before operation showing the intracardiac tumour filling the inferior vena cava, the right atrium and extending into the right ventricle. (The arrow points to the tumour and tumour thrombus).

After pre-operative preparation, the child underwent radical resection of nephroblastoma + incision and thrombectomy of inferior vena cava + removal of tumour thrombus in right atrium + implantation of venous infusion port. During the operation, the right kidney with tumour was completely resected, the perirenal fat was removed, and the right adrenal gland was preserved. The right anterior and lateral wall of the inferior vena cava was cut longitudinally for about 10 cm. The tumour thrombus was solid and brittle, closely adhered to the wall of the inferior vena cava. It was about 10 cm long and uneven in diameter. The widest part was about 1.5 cm. The wall of the inferior vena cava was repaired with sutures. The tumour thrombus entered the right atrium through the inferior vena cava, and the head of the tumour thrombus was obviously enlarged, with a transverse diameter of about 6 cm and a total length of about 17 cm. Obvious adhesions made it impossible to pull out from the inferior vena cava, so we decided to perform right atrial tumour thrombus removal with cardiopulmonary bypass support. The median incision was made in the anterior chest, after heparinisation, the aorta and the superior vena cava were intubated in sequence. After the temperature was lowered and the superior vena cava was blocked, the right atrium was cut to expose the tumour thrombus. The adhesion of the tumour thrombus to the right atrium and inferior vena cava was carefully stripped, the tumour thrombus tissue was removed from the atrium, the remaining tumour thrombus was basically completely removed, the inferior vena cava backflow was smooth, the right atrium incision was sutured, and the right atrium incision was sutured after warming up. The heart did not stop beating during the operation (Fig. 2).

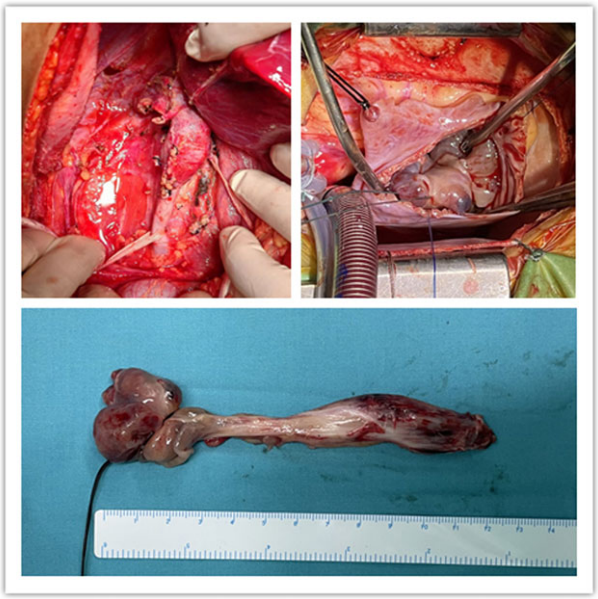

Figure 2. Complete removal of tumour thrombus in right atrium and inferior vena cava.

The child returned to the ICU after surgery. Supportive symptomatic treatment was given after the operation. In order to prevent venous thrombosis, heparin sodium was continuously intravenous infusion after the operation to maintain the APTT at about 1–1.5 times the normal value. The endotracheal tube was extubated 1 day after the operation, and the child transferred out of the ICU 2 days after the operation. Post-operative thrombosis was not seen, so we changed to oral aspirin anticoagulant therapy 7 days after the operation. Post-operative echocardiography showed the inner diameters of the right atrium and right ventricle were within the upper limits of normal, and no tricuspid regurgitation was seen.

The child was discharged 14 days after the operation and returned to the local hospital to continue chemotherapy. No tumour recurrence, no tumour thrombus, and thrombus growth were found in the 6-month follow-up examination of the child, and echocardiography showed minor tricuspid regurgitation, and no special treatment required.

Discussion

Cardiac mass in children is rare, with insidious onset, and primary cardiac mass is less than secondary mass. However, with the popularisation of prenatal examination and deepening understanding of such diseases, more attention has been paid to heart space-occupying diseases. According to literature reports, about 4–10% of children with Wilms tumour are accompanied by tumour thrombus in the venous system.[4,5] Among the malignant tumours in children with tumour thrombus in the venous system, about 98% of the cases are nephroblastoma,[6,7] but it is still rare for the tumour thrombus to reach the level of the atrium or even enter the right ventricle.

In this case, the child had an insidious onset. He came to the doctor with palpitation and chest tightness. He had already developed symptoms related to the circulatory system. The joint examination of the chest and abdomen revealed a space occupying

https://doi.org/10.1017/S1047951123002652 Published online by Cambridge University Press

kidney. The insidious onset and rapid progression were consistent with the pathophysiological characteristics of Wilms tumour. Perioperative chemotherapy can reduce surgical trauma, and some patients with intracardiac tumour thrombus or inferior vena cava tumour thrombus may avoid thoracotomy to remove tumour thrombus and reduce the scope of surgery. However, after pre-operative chemotherapy, although the kidney tumour shrank in this child, the venous tumour thrombus still filled the inferior vena cava, reached the right atrium, and protruded to the right ventricle, so the classification of the venous tumour thrombus still belonged to type IV. Surgical is the only way to treat the kidney tumour and the cardiac tumour thrombus. Abdominal incision was used to separate the tumour thrombus in the inferior vena cava. After thoracotomy, the aorta and superior vena cava were catheterised to establish extracorporeal circulation without cardiac arrest. After checking the unobstructed blood flow of the inferior vena cava, the atrium was sutured. This is different from the approach of Abdullah et al.[8] They used cardiopulmonary bypass with deep hypothermia and circulatory arrest to remove the intracardiac tumour. We believe that that surgical method prolongs the time of extracorporeal circulation and causes significant interference and damage to the organs. Therefore, we adopted the cardiopulmonary bypass bypass method to remove the tumour thrombus as soon as possible. Surgical operations minimise trauma and remove the tumour thrombus as completely as possible. However, if the tumour thrombus is significantly adhered to the surrounding area, part of the tumour thrombus can be left to ensure the integrity of blood vessels and the heart. Current studies have shown that residual tumour thrombus is beneficial for Wilms tumour patients. Upstaging in case of incomplete resection with additional radiotherapy after surgery may overcome a negative impact on survival after incomplete resection of VCT.[9]

According to statistics, the incidence of venous thrombosis in patients with malignant tumours is 4–7 times that of healthy people,[10] and about 20% of newly formed venous thrombosis is closely related to the occurrence of tumours.[11] Patients with malignant tumours are usually in a state of blood hypercoagulability or prothrombotic state, and the blood is abnormal in one or more links of the Virchow triangle (venous system endothelial cell injury, venous blood stasis, and blood hypercoagulability) and the immune system. This leads to deep vein thrombosis.[12,13] Therefore, the child was given routine anticoagulant therapy after the operation, and the changes in coagulation function were monitored to maintain the APTT at 1–1.5 times the normal value. The post-operative re-examination of the child showed no thrombosis, and the child was given oral aspirin for anticoagulant therapy at 7th day after the operation. Six months later, the child's review showed no thrombus formation. Although the incidence of intracardiac space-occupying from nephroblastoma is low, in recent years, multi-centre reports of accidents related to tumour thrombus, such as pulmonary embolism, cardiac arrest, atrial obstruction.[14–16] Pre-operative radiotherapy and chemotherapy although some patients with tumour thrombus can be significantly shrunk to avoid thoracotomy, if the tumour thrombus is not sensitive to this, surgical treatment should be combined as soon as possible to avoid the risk of circulatory system. Just like what we have done to this child.

Conclusion

Paediatric cardiac tumour thrombus is a serious condition that requires prompt and effective management to prevent potentially fatal outcomes. The appropriate treatment will depend on the size and location of the tumour, as well as the overall health status of the child. Conservative treatment with anticoagulant medications may be appropriate in some cases, while surgical intervention may be necessary in others. Regardless of the approach chosen, it is important that the condition is managed by a team of experienced healthcare professionals to ensure the best possible outcomes for the child.

Competing interests. The authors declare no competing interests.

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https://doi.org/10.1017/S1047951123002652 Published online by Cambridge University Press