Cardiology in the Young

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Kaposiform hemangioendothelioma of the heart: a case report and literature review

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Brief Report

Cite this article: Fan D, Cui Y, Chen J, and He X (2023) Kaposiform hemangioendothelioma of the heart: a case report and literature review. *Cardiology in the Young* **33**: 2411–2414. doi: 10.1017/S1047951123001269

Received: 12 November 2022
Revised: 27 April 2023
Accepted: 4 May 2023
First published online: 19 June 2023

Keywords:
Kaposiform hemangioendothelioma; echocardiography; pericardial effusion

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Abstract

Kaposiform hemangioendothelioma is a rare tumour of vascular origin that rarely occurs in the heart. We provided a rare case of a 26-day-old infant with tachypnoea. Echocardiography showed a solid tumour in the pericardial cavity and a large amount of pericardial effusion. The solid tumour was confirmed by surgery, and the pathology was kaposiform hemangioendothelioma. We analysed this case and reviewed the related literature to explore the clinical features and echocardiographic manifestations to improve the understanding, diagnosis, and treatment of this disease for clinicians and sonographers.

Kaposiform hemangioendothelioma is a kind of borderline vascular tumour that rarely occurs in the heart; only three cases have been reported.[1–3] Furthermore, ultrasonography features of cardiac kaposiform hemangioendothelioma have rarely been reported. Herein, we report a rare case, wherein echocardiography showed an extensive solid tumour in the pericardial cavity, without extension into a cardiac chamber, whilst surgery demonstrated extensive myocardial invasion. The related literature was reviewed to summarise the clinical features and echocardiographic manifestations.

Case data

Case report

The sick 26-day-old girl was admitted to the hospital 24 days after a pericardial effusion was found. During pregnancy, ultrasonography demonstrated a foetal pericardial effusion. Two days after birth, echocardiography in the birth hospital revealed a patent foramen ovale, patent ductus arteriosus, and a large pericardial effusion. After treatment, she improved and was discharged from hospital. Twenty-six days after birth, echocardiography in the local hospital showed a large pericardial effusion again. The child was transferred to our hospital for further diagnostic imaging and treatment. Physical examination showed shortness of breath with dyspnoea, nasal flaring, and subcostal recession. Initial echocardiography showed a patent ductus arteriosus, patent foramen ovale, and large pericardial effusion. Pathocytological examination of pericardiocentesis fluid showed hyperplastic mesothelial cells, a few monocytes and lymphocytes, and many red blood cells were found on smear. Echocardiography after the pericardial effusion was relieved showing that there was an extensive solid tumour in the pericardial cavity, extending from the top of the right atrium to the top of the left atrium. It extended leftward and downward along the posterior wall of the left atrium to the posterior atrioventricular groove, surrounding and compressing the coronary sinus(Fig. 1). Ultrasonic examination of the tumour suggested a possible pericardial haemangioma. There was no decreasing trend in the pericardial effusion after intermittent pericardiocentesis. Given the persistent reaccumulation of the pericardial effusion and that it was related to a solid tumour in the pericardial cavity with failed conservative treatment, she was transferred to the cardiac surgery unit for surgical treatment. During the operation, the ventricle was seriously infiltrated by the tumour (Fig. 2) and had invaded the myocardium, so it could not be removed completely. Only a small part of the tumour tissue located at the top of the right atrium was resected and sent for pathological examination. Post-operative histopathology revealed a kaposiform hemangioendothelioma, with tumour tissue invaded the surrounding striated muscle.

Literature review

In Chinese, "Kaposiform hemangioendothelioma" was used as the search term in the China National Knowledge Infrastructure (CNKI) and Wanfang databases, and in English, "Kaposiform hemangioendothelioma" was used as the search term in the METSTR and PubMed databases to search the related literature of cardiac kaposiform hemangioendothelioma, which was reported from the establishment of the database to July 2022. Three English literature case reports were found, giving, together with this case, data on four sick infants (Table 1). All cases were confirmed by surgery and pathology.

Table 1. General situation, clinical symptoms, echocardiographic manifestations, and treatment of four children with cardiac kaposiform hemangioendothelioma[1-3]

Serial number	Date	Gender	Age	Main clinical symptom	Kasabach–Merritt phenomenon	Echocardiographic manifestations	Treatment and prognosis
One	2008	Male	1 day	Progressive wheezing	Positive	Pericardial effusion. Homogenous mass at the base of heart surrounded the aorta and pulmonary artery.	Tumour only partially resectable. Vincristine combined with steroids used for treatment with significant tumour size reduction by age 6.5 months. Tumour not fully resectable with no further treatment. After 6 months, no obvious growth of the tumour. Tumour not fully resectable. Vincristine combined with steroids used with no obvious improvement., Sirolimus added with significant size reduction after six months.
Two	2013	Male	2 months	Dyspnoea	Negative	Large pericardial effusion with large homogeneous mass posterior to left atrium, extending to intracardiac cavity.	
Three	2018	Male	24 days	Dyspnoea	Positive	Pericardial effusion and isolated thickened of the left atrium with homogenous echogenicity of its roof, lateral wall, and the interventricular septum.	
Four	2022	Female	26 days	Tachypnoea	Negative	Large pericardial effusion. Solid tumour in pericardium with coronary sinus compression.	Tumour not fully resectable. Sirolimus used with significant tumour size reduction after 6 months, with coronary sinus returning to normal, and pericardial effusion resolved

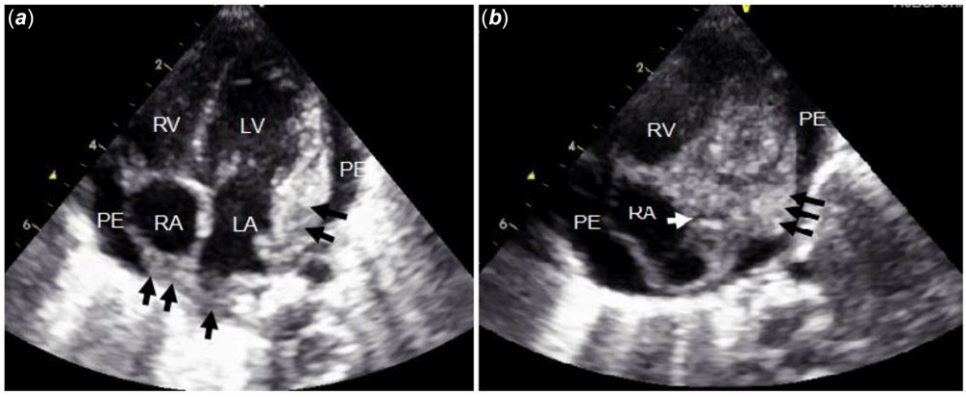

Figure 1. ***a*** Apical four-chamber echocardiographic view showing solid tumour in pericardial cavity, extending from the top of the right and left atriums to the rear wall of the left atrium and the posterior atrioventricular groove (black arrow), accompanied by pericardial effusion. ***b*** Posterior four-chamber echocardiographic view showing solid tumour (more obvious than the apical four-chamber plane). The boundary with the myocardium is unclear (black arrow), surrounding and compressing the coronary sinus (white arrow). Note: LA left atrium, RA right atrium, LV left ventricle, RV right ventricle, PE pericardial effusion.

Discussion

Kaposiform hemangioendothelioma is a kind of borderline vascular tumour of endothelial origin with local invasion[4] that has the dual characteristics of haemangioma and Kaposi's sarcoma.[5] In 1993, Zukerberg[6] and others first described and coined the term "kaposiform hemangioendothelioma." In 2014, the International Society for the Study of Vascular Abnormality (ISSVA) classified it as a "locally invasive or borderline vascular tumour."[7]

Kaposiform hemangioendothelioma is very rare but when diagnosed, it is usually in infants and children.[8] This disease often occurs in the skin and deep soft tissues of the limbs, upper torso, and maxillofacial region.[9] This case occurred in the heart, a rare manifestation, and recently has been successfully treated by surgery and sirolimus. The pathogenesis of this disease is still unclear, and it may be related to chromosome abnormalities.[10] The main clinical manifestations of cardiac kaposiform hemangioendothelioma are dyspnoea and tachypnoea. Kaposiform hemangioendothelioma grows, and surrounding tissues of the tumour are often invaded. It does not regress spontaneously, and it may be accompanied by regional lymph node metastases, but distant metastatic spread is rare, with only one case reported.[11,12]

Kaposiform hemangioendothelioma is often non-specific on laboratory investigations. If it is accompanied by the Kasabach–Merritt phenomenon, there may be some abnormalities, such as thrombocytopenia, fibrinogen decrease, and D-dimer increase. Studies have shown that approximately 71% of kaposiform hemangioendotheliomas can develop into Kasabach–Merritt syndrome. In this study, two of the four children were accompanied by the Kasabach–Merritt phenomenon.[1–3] According to statistics, the Kasabach–Merritt phenomenon is the main cause of death in sick children, with mortality rate up to 30%.[5,13]

Cardiac tumours are rare in newborns and are easily missed and or misdiagnosed. The child's first echocardiographic examination in our hospital and the local hospital did not find a cardiac tumour.

https://doi.org/10.1017/S1047951123001269 Published online by Cambridge University Press

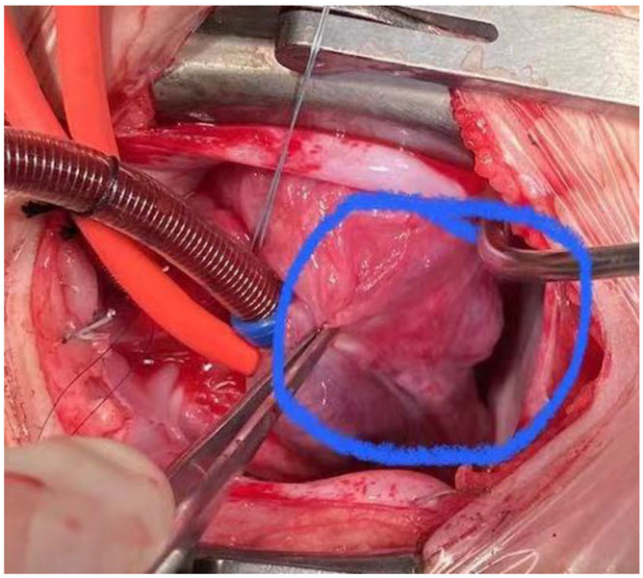

Figure 2. During the operation, the tumour was located at the back of the heart and surrounding the coronary sinus, with serious infiltration and unclear boundary with the myocardium (in the blue area).

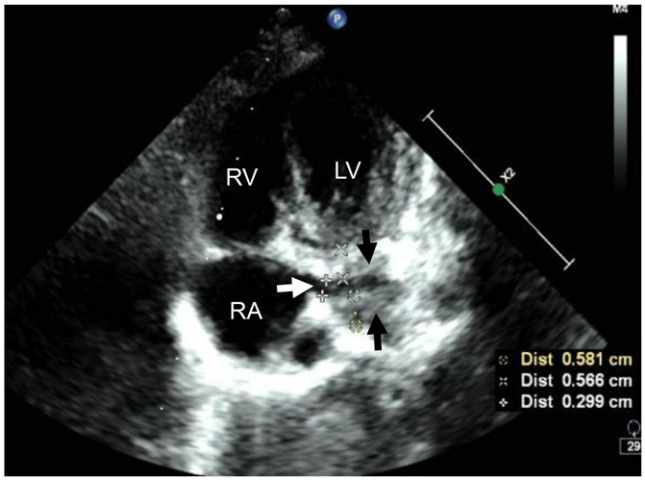

Figure 3. After 6 months, the tumour had significantly reduced in size (black arrow), the size of the coronary sinus had returned to normal (white arrow), and pericardial effusion had disappeared. LV = left ventricle; RA = right atrium; RV = right ventricle.

In this case, the difficulty of ultrasound scan was related to conventional echocardiographic section and their inability to show the tumour because it was located at the top of the atriums and ran along the posterior wall of the left atrium, extending to the coronary sinus and the bottom of the right ventricle, with a wide range and invasive growth. The conventional apical four-chamber cutting plane did not easily display the tumour. When the sound beam was tilted backward in an "unconventional" plane to show the coronary sinus (the posterior four-chamber of heart), the tumour located behind the heart could be shown. During the examination, the doctor only scaned conventional sections, focusing on the large pericardial effusion and cardiac function, thus not identifying tumour located behind the heart, resulting in a missed diagnosis. We now encourage our sonographers to pay attention to both routine standard section scanning and unconventional section scanning in their daily practice, especially in the setting of an unexplained pericardial effusion, as here. Cardiac kaposiform hemangioendothelioma occurs mainly in the pericardium, and it is rare to extend into the intracardiac cavity, although some can invade the myocardium. Echocardiography can be relied upon to show a solid tumour within a large pericardial effusion, whether located in the pericardial cavity or around the heart invading the pericardium, as well as pericardial thickening with nodular protrusion. However, this may be challenging if the tumour is diffuse, without a clear boundary with surrounding structures. If it is accompanied by the Kasabach–Merritt phenomenon, kaposiform hemangioendothelioma should be considered by ultrasonic examination.

The invasive and progressive growth of kaposiform hemangioendothelioma is different from that of typical infantile haemangioma, as the latter has a proliferative phase and then regresses.[14] Echocardiography of infantile haemangiomas shows that they are often localised and can be nodular; in contrast, kaposiform hemangioendotheliomas tend to infiltrate diffusely with ill-defined borders. The definite diagnosis depends on pathological examination.

Cardiac kaposiform hemangioendothelioma presents with severe symptoms and, once found, should be treated agressively. Surgical resection is the first choice, whilst for those that cannot be removed completely by surgery, vincristine combined with steroids has been effective. If conventional treatment is ineffective, one study reports successful tumour regression with sirolimus on a clinically refractory case.[15] When sirolimus is used, it is very important to use prophylactic antibiotics, monitoring drug levels regularly. In our case, because the tumour invaded the myocardium, complete operative removal was not possible, and the patient was treated with sirolimus for half a year. At present, good results had been achieved in short-term follow-up: the tumour has clearly shrunk in size, with coronary sinus diameter returning to normal, and the pericardial effusion disappeared (Fig. 3).

Conclusion

Cardiac kaposiform hemangioendothelioma is a rare cardiac space-occupying lesion, with some cases accompanied by the Kasabach–Merritt phenomenon. It is more common in newborns and infants. It often occurs in the pericardium and is often located at the back of the heart, with a wide range of invasive myocardial growth, which can extend into a cardiac chamber. It may be accompanied by a large and persistent pericardial effusion. Echocardiography, as a real-time, non-invasive, and convenient imaging examination, is the first choice for diagnosis with specific imaging characteristics. The timely detection of the disease is very important for the treatment and prognosis of sick infants.

Acknowledgements. I would like to express my gratitude to all those who helped me during the writing of this thesis.

Author contribution. Di Fan wrote the main manuscript text.

Yun Cui and Jing Chen contributed to the interpretation.

Xinjian He provided echocardiographic expertise and perspective for the case summaries and discussion.

Financial support. This research received no specific grant from any funding agency, commercial, or not-for-profit sectors.

Competing interests. None.

https://doi.org/10.1017/S1047951123001269 Published online by Cambridge University Press

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