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A RARE CASE STUDY: A MULTIPLE MYXOMAS INVOLVING BOTH RIGHT AND LEFT VENTRICLES IN NEONATE

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ABSTRACT
Cardiac myxomas are rare, with multiple myxomas even rarer (0.001%–0.03% incidence). They primarily arise in the left atrium (75%), with 20% in the right atrium, and less than 5% in the ventricles. Multiple ventricular myxomas without atrial involvement is extremely rare. Myxomas are mostly benign, and advances in imaging have made them more manageable. This case discusses a neonate with multiple myxomas in both ventricles. A 29-week preterm neonate, post-C-section, was admitted to the NICU with signs of pneumonia. Routine echocardiography revealed multiple masses in both ventricles. The right ventricle had five masses (ranging from 0.3–1.49 cm), and the left ventricle had three masses (ranging from 0.2–0.7 cm). Cardiac myxomas are commonly atrial. Atrial myxomas causing obstruction, thromboembolisms, and constitutional symptoms, while ventricular ones may cause chest pain, palpitations, or syncope. Diagnosis is made via histopathology and echocardiography. This patient presented with multiple mobile myxomas in both ventricles. Cardiac myxomas are rare tumors, with low incidence, typically affecting atrium, while multiple myxomas involving both ventricles are rare. This case describes a premature neonate diagnosed with multiple myxomas in both ventricles, exhibiting mild respiratory distress that improved with CPAP. Despite the presence of the tumors, there were no significant symptoms. Surgical resection is the standard treatment, with a low mortality rate and favorable recovery. Early detection via imaging and surgical intervention are crucial for optimal outcomes.

KEYWORDS: Cardiac tumor, myxoma, left ventricular myxoma, right ventricular myxoma, echocardiography

INTRODUCTION

Myxoma is one of the most common heart tumour, although overall heart tumour is one of the tumour with the lowest incidence rate. Where it is estimated that there are only 0.5 cases out of 1 million population per year (Shohei K. 2016). Generally, if more than 1 tumour is found in the human heart, it shows the presence of malignancies or is the result of metastasis. Most of the malignant heart tumour is sarcoma-like *angiosarcoma, rhabdomyosarcoma, fibrosarcoma,* and *leiomyosarcoma*. This type of heart tumour has a poor prognosis both for treatment with medication and with surgery (Shohei K. 2016). Myxoma is mainly the most common type, atrial myxoma can be found with three main symptoms or complications, namely duct obstruction, embolism and constitutional symptoms such as fever and weight loss (Pinede L. 2001).

Based on immunohistopathological findings, myxoma cells were found to be derived from multipotent mesenchymal cells, where the cells have the ability to become neural and endothelial cells (Pucci A. 2000). Epidemiologically, most myxomas are found in female patients with a peak incidence in the fourth and sixth decades (Yu K. 2007) (Wu HM. 2019). There are several studies that have found an estimated ratio of myxoma incidence in women and men to be 2.05 : 1 and 0.75 : 1 for the left and right atriums. Very few cases of myxoma have been found in pediatric patients (Yu K. 2007) (Onubogu U. 2017)

There are 3 clinical symptoms that are commonly found in patients with multiple myxomas, these symptoms can appear through various mechanisms. Of the 3 clinical symptoms, obstruction and thromboembolic have different manifestations based on the myxoma location, while the constitutional symptoms are not affected by the myxoma location. There are several other symptoms but these symptoms can be linked to the carney complex, so these symptoms are not discussed deeply in this case report. This case report will discuss multiple myxoma without carney complex in neonate patients.

RESEARCH METHODS

The research method used in this study was a qualitative approach employing a case study design, focusing on an in-depth exploration of the rare phenomenon of multiple myxoma in neonates, as described in the document. The research subject was a single premature neonate patient admitted to the NICU, with data collection techniques involving direct clinical observation, medical record documentation, and results from supportive examinations such as echocardiography, which were used to identify the tumor's characteristics and location. Data were analyzed using

qualitative descriptive methods by interpreting clinical findings, comparing them with relevant scientific literature, and comprehensively assessing the patient's condition to understand the characteristics, symptoms, and management of multiple myxoma in neonates. Data validity was strengthened through source triangulation—the consistency between observational results, medical data, and scientific references—thereby yielding a deep and contextual understanding of the cases studied.

Case report

A neonate is taken from the operating room after *a Sectio Caesaria* to the NICU with a preterm birth at 29 weeks 1 day; that neonate has sufficient crying and movement, along with vital signs and general status which are still in a normal level. Neonates undergo echocardiography as a routine examination of congenital heart disease, mainly because the patient has a premature status. From the echocardiography examination it was found that the site of the solitary atrium, systemic and pulmonary veins were normal, *AV-VA* concordant, four balanced chambers, 1.3 mm LA/AO, no *PFO,* no *ASD*, no *VSD*, no *PDA*, no *CoA*, left aortic arch, *TR* mild, no pericardial effusion, and normal left ventricular systolic function (EF: 68%). Several masses are visible in the right ventricle: I: 1.49 x 1.04 cm, II: 0.3 x 0.32 cm, III: 0.5 x 0.3 cm, IV: 0.3 x 0.49 cm, V: 0.4 x 0.47 cm; in the left ventricle: I: 0.7 x 0.4 cm, II: 0.2 x 0.2 cm, III: 0.5 x 0.4 cm. The patient does not show symptoms of multiple myxoma; There is no desaturation and only had a slight tachypnea, hypothermia, and very minimal chest retraction. The patient is currently awaiting surgical intervention from a cardiothoracic surgeon and histopathological examination. There is no history of multiple myxoma from the other siblings, and the sibling was born without myxoma or other congenital heart disease.

DISCUSSION

Cardiac myxoma is the most common type of heart tumour. Most cardiac myxomas are found in the atrium, especially the left atrium (Schaff W. 2000). The incidence rate of cardiac myxoma is higher in women than men. Myxoma in the ventricles is very rare, especially myxoma that includes the left and right ventricles (Bakaeen FG. 2003). There are some differences from myxoma found in the atrium and ventricle, where in the left atrial myxoma, the myxoma is most commonly found in the mitral annulus or in the oval fossa of the interatrial septum. Whereas in ventricular myxoma, myxoma can be found in all parts of the ventricles. From a study by Abad C et al, it was found that 24 out of 71 cases of left ventricular myxoma came from the ventricular septum and 46 out of 71 came from the left ventricular wall (Abad C. 2014). This causes ventricular myxoma to become more mobile, which causes risk of many symptoms such as embolism, arrhythmia, blockage of the outflow tract and other constitutional problems (Robert J. 2009) (Arruda MV. 2008). In this case, the patient was a female neonate, in whom a myxoma was found in the right and left ventricles. There are multiple myxomas where most of which are found in the ventricular wall.

There are 3 symptoms that are commonly found in patients with atrial myxoma, namely, obstructive, thromboembolic and constitutional symptoms (Goswami KC 1998) (Wang Z. 2016) (MA G. 2019). However, for symptoms in myxoma involving the ventricles due to limited data, it is still not possible to conclude with certainty the general symptoms that can represent ventricular myxoma. Some literature has found that for ventricular myxoma the symptoms that arise are closely related to the location where the myxoma is found. Chest pain, palpitations, syncope, systemic embolism and complications from such symptoms such as arrhythmia to sudden death, can be symptoms that can be found in ventricular myxoma (Xia LY. 2002). In that patient, no specific symptoms were found, and the symptoms that could be found were only hard breathing and chest retraction, which were very minimal and immediately improved after being given oxygen.

Histopathological examination is still the gold standard to determine the exact diagnosis of myxoma, where in the results of the pathological examination findings can be found in the irregular lesions with spindles, brittle with an indented capsule. Meanwhile, in histological examination, myxoma has irregular or star-shaped cell characteristics that spread to mucoid cells (Ji X. 2021). However, in this patient, a histopathological examination has not been done.

On physical examination, it is generally possible to find abnormalities through *a tumour flop* that can be heard in the early diastolic phase, but this symptom is more commonly found in patients with myxoma in the left atrium (Bakaeen FG. 2003). As for the findings from physical examination in patients with ventricular myxoma more towards complications than other symptoms, because the symptoms of ventricular myxoma are highly dependent on the location of the myxoma itself, various symptoms can be found such as, *dysnea on effort*, hypoxia, tachycardia, blindness, transient ischemic attack and various neurological defects can also be found such as hemiplegia and even sudden death can happen (Lee VH 2007).

Echocardiography *is* still an option to get an early diagnosis, this is because echocardiography tests are noninvasive, safe for patients and accurate. The echocardiography findings on myxoma are made through the description of the hyperechoic period in the ventricles, where the period can move following the contraction and relaxation of the heart (Ji X. 2021). However, this echocardiography picture must be distinguished from the picture of the thrombus, where when compared to the thrombus, the myxoma tends to be more mobile and has a narrow base and wide pedicle, while the thrombus has a solitary period, seemingly dense, homogeneous solid with a size from 2 to 10cm (21). This is found in patients where there is a hyperechoic multiple period and seems mobile, the multiple period can be observed in the

right ventricle: I: 1.49 x 1.04 cm, II: 0.3 x 0.32 cm, III: 0.5 x 0.3 cm, IV: 0.3x0.49 cm, V: 0.4 x 0.47 cm, and in the left ventricle: I: 0.7 x 0.4 cm, II: 0.2 x 0.2 cm, III: 0.5 x 0.4 cm. In addition to echocardiography examinations, CT scans and MRI can be the alternative if the results of echocardiography are inconclusive, but in this patient because the echocardiography is conclusive, CT scan and MRI examinations were not made.

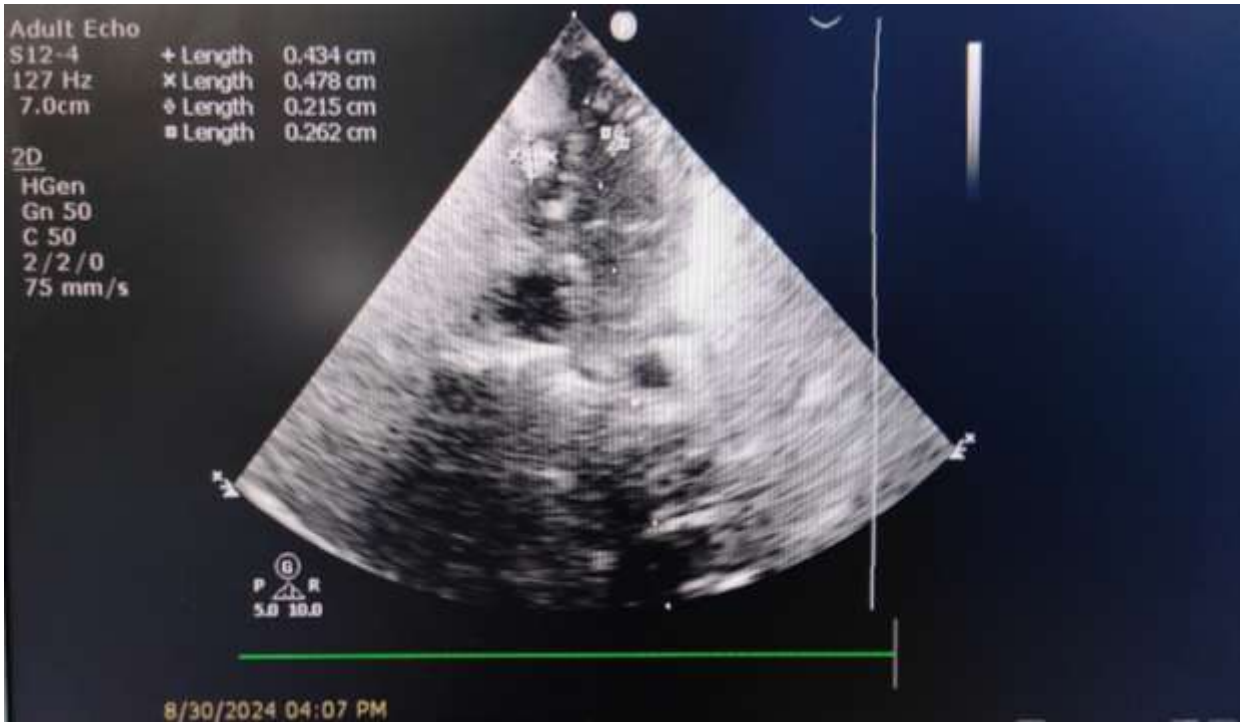

Image 1. Apical 4 chambers view RV and LV

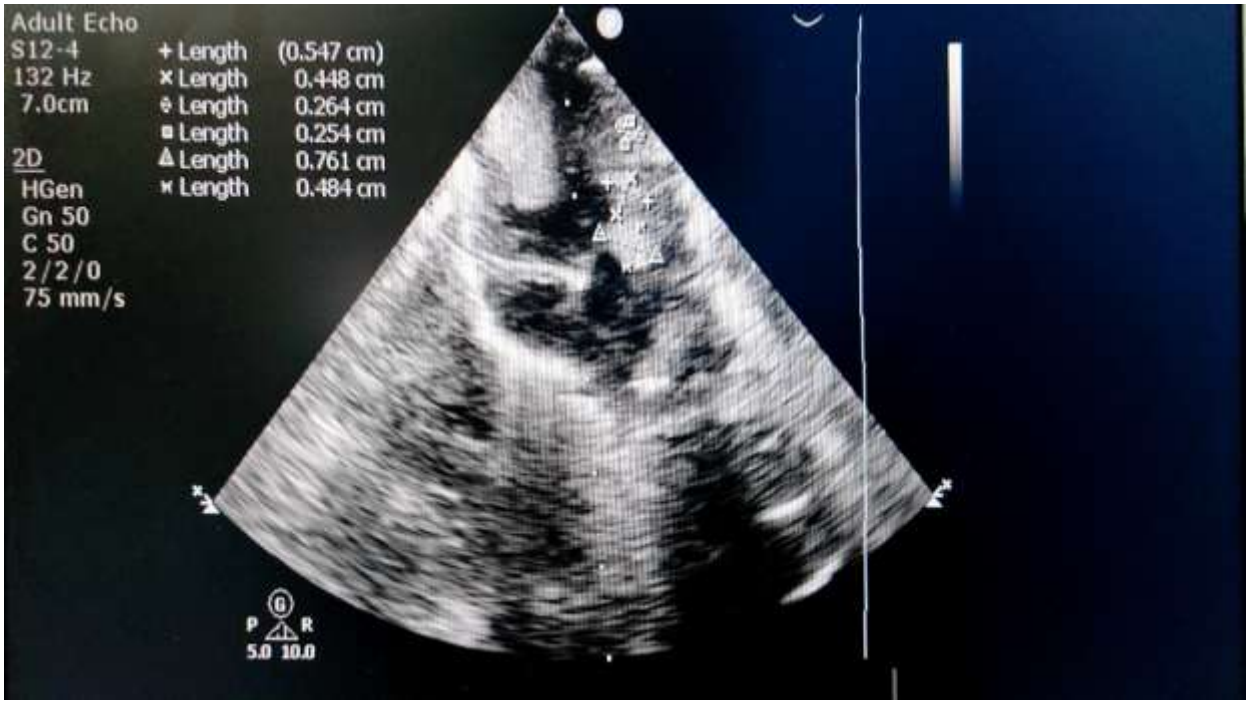

Image 2. Apical 4 chambers view LV

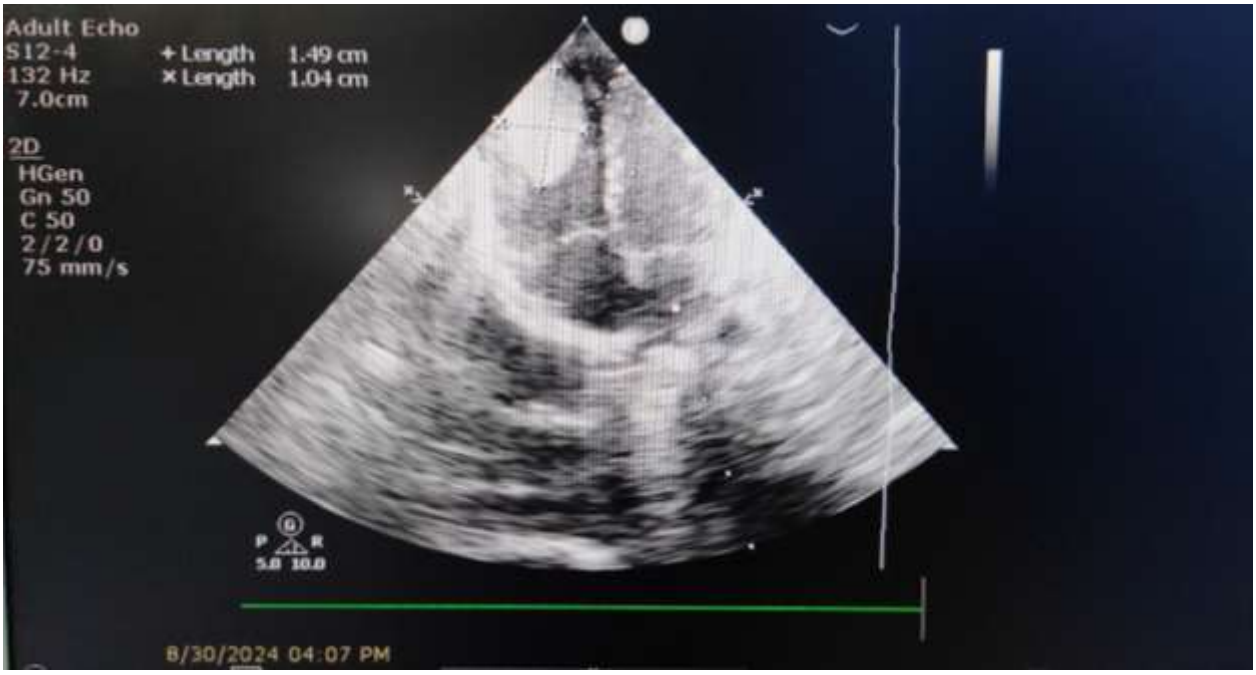

Image 3. Apical 4 chambers view RV

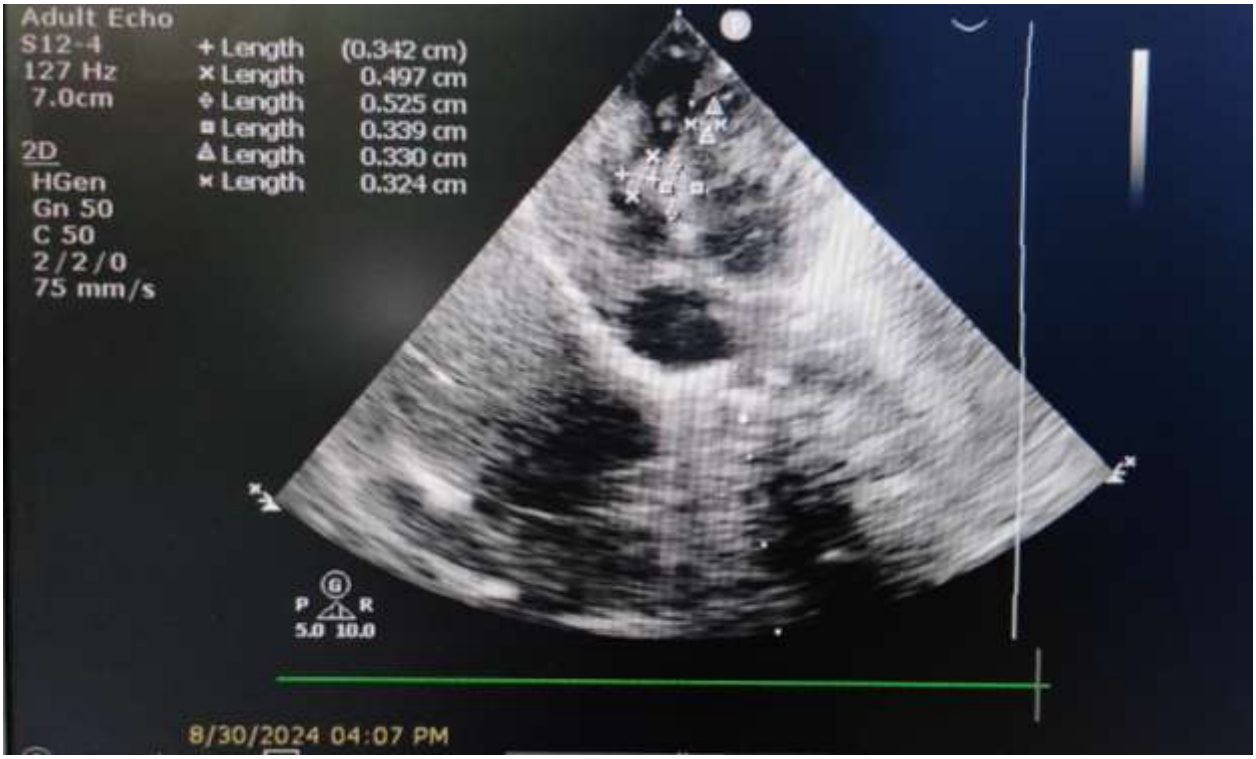

Image 4. Apical 4 chambers view RV 2

Myxoma can develop up to 0.15 cm each month, although there are no symptoms or blockages or embolism, operative action should be taken immediately after the patient is diagnosed (Qin W. 2014). Complete resection operative action is still the main choice for myxoma, but myxoma recurrence will happen if the resection is not completely successful (Kim HY, 2012). The prognosis of this resection procedure is also excellent where the mortality rate does not reach 5% with rapid postoperative recovery, there are several complications that can occur although rarely found in this resection procedure such as ventricular dysfunction, atherosclerosis, respiratory distress and infection but with the risk of not performing heavier operative measures which until now operative action is still an option (Liu L. 2020). If operative action is not taken immediately, various complications can arise, such as symptoms of congestive heart

failure, arrhythmias, valvular defects, thromboembolic and infections can also happen. Currently, the patient is waiting for a schedule for operative action and histopathological examination at RSUP Prof. Ngoerah Bali.

CONCLUSION

In conclusion, cardiac myxoma is a relatively common heart tumour, although heart tumours themselves are very rare. Cardiac myxoma involving the left and right ventricles is a very rare case. Regular examinations of neonates showing symptoms of cardiac myxoma with basic echocardiography can be a life-saving procedure or at least can reduce the likelihood of very serious complications for the patient. Although most multiple myxomas are associated with Carney's complex, which is a genetic condition, in this case, the complex is not found in the patient, or may not have developed due to the patient's age. The use of multi-modality radiology, such as echocardiography and CT angiography along with histopathology may be necessary to make an accurate diagnosis, and surgical intervention should be performed as soon as the diagnosis is confirmed. The prognosis for patients undergoing surgical resection of myxoma is impressive. The death risk during surgery is less than 5 percent, and patients usually recover quickly after surgery. The chance of tumour recurrence is 1% to 3% in sporadic cases, 12% in familial cases, and 22% in complex myxoma. One study showed that minimal manipulation of the tumour, excision with sufficient margins, and a thorough examination of all the heart chambers are important steps to prevent tumour from appearing again. With advances in cardiac imaging, myxoma can now be detected earlier and treated. Until a few decades ago, most myxoma cases were only found on autopsy.

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