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Case Report

Patient with myelodysplastic syndrome presented with recurrent pericardial effusion diagnosed as epicardial hemangioma; Case report of a rare diagnosis with rare presentation ☆☆☆

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ABSTRACT

Primary cardiac tumors are extremely rare and cardiac hemangiomas comprise less than 3% of them. Presentation of such disease with recurrent pericardial effusion is even rarer. Our patient is known case of myelodysplastic syndrome and up to our knowledge there are no reported case in which cardiac hemangioma was diagnosed in a patient with myelodysplastic syndrome. This 64 years male patient presented to our department with recurrent pericardial effusion, diagnosis was a query after extensive work he was found to suffer from a cardiac tumor based on the pulmonary artery and right ventricle. We performed surgery for him on cardiopulmonary bypass and did complete resection of the mass for him and result of biopsy showed mixed hemangioma. Recurrent pericardial effusion is most commonly a sign of a malignancy. Even with advancement of medical technology diagnoses of

Abbreviations: MDS, myelodysplastic syndrome; CT, computed tomography; MRI, magnetic resonance imaging.

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cardiac hemangiomas are still difficult. Treatment and definitive diagnosis is still complete surgical resection and histopathological examination.

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Introduction

Primary cardiac tumors are extremely rare [1,2]. The majority of studies are based on autopsy findings, which comprise 17 in a million to 0.28 per cent of autopsy cases [1,2]. Secondary metastasis, on the other hand, is much more common [2]. Approximately 75% of these primary tumors are benign, and most of these are myxomas, which constitute 50% of all primary cardiac tumors [2]. Cardiac hemangiomas are extremely rare, accounting for less than 3% of all primary cardiac tumors [1,3]. In addition, cardiac hemangiomas presenting with effusion or tamponade are extremely rare [1,4]. Myelodysplastic Syndrome (MDS) is a clonal marrow stem-cell disorder characterized by ineffective haemopoiesis resulting in anemia [5]. The progression of acute myeloid leukemia has been reported in several cases of patients [5]. This report presents the case of a patient with recurrent pericardial effusion who, upon workup, was found to have a mass on the epicardium that was subsequently resected.

Case report

The patient, a 64-year-old man with a history of MDS, was referred to the Cardiac Surgery Department in Sulaymaniyah due to recurring pericardial effusions.

It began 1 year earlier when he was diagnosed with pericardial effusion and had dyspnea and orthopnea. After he failed to respond to medical therapy, he underwent pericardiocente-

sis, and 1 L of serous fluid was extracted. A cytological examination of the sample revealed no evidence of malignancy.

After 3 months, the patient developed dyspnea and chest pain and was diagnosed with recurrent pericardial effusions. No diagnosis was found after multiple imaging and cytological examinations of the fluid after re-aspiration.

The patient was referred to our department with signs of yet another pericardial effusion, and we began a diagnostic workup.

Besides MDS, for which he is receiving Danazol treatment and repeated transfusions when hemoglobin levels fall below 8, he has no history of other chronic ailments. He has been put on Danazol treatment for the last 4 years because he developed thrombocytopenia. He receives no other form of treatment.

He does not smoke or consume alcohol regularly. A physical examination revealed that the patient is dyspnoeic and cannot lie comfortably.

A transthoracic echocardiogram (using Philips CX50 Ultrasound Machine and a linear transducer) revealed a hypoechoic mass near the pulmonary valve on the right side of the heart (Fig. 1). Chest X-Ray showed cardiomegaly (Fig. 2). CT scan of the chest with IV contrast (dilute iodinated contrast) revealed a 19 mm outpouching on the left of the pulmonary trunk, and the structure showed central isodensity. In addition, there was a large amount of pericardial effusion (Figs. 3A and B).

Surgical exploration was decided upon. A classical sternotomy was performed in which the pericardium was opened, a large quantity of fluid was drained from the pericardium, and a tissue sample was taken for cytology. The right ventric-



Fig. 1 – Transthoracic echocardiography showing a mass in right side of heart (arrow) and pericardial effusion.

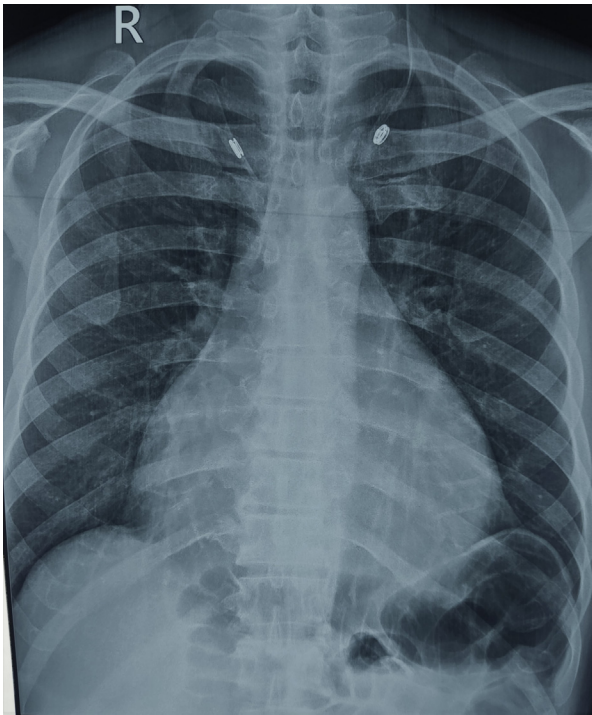


Fig. 2 – Chest X-ray showing cardiomegaly.

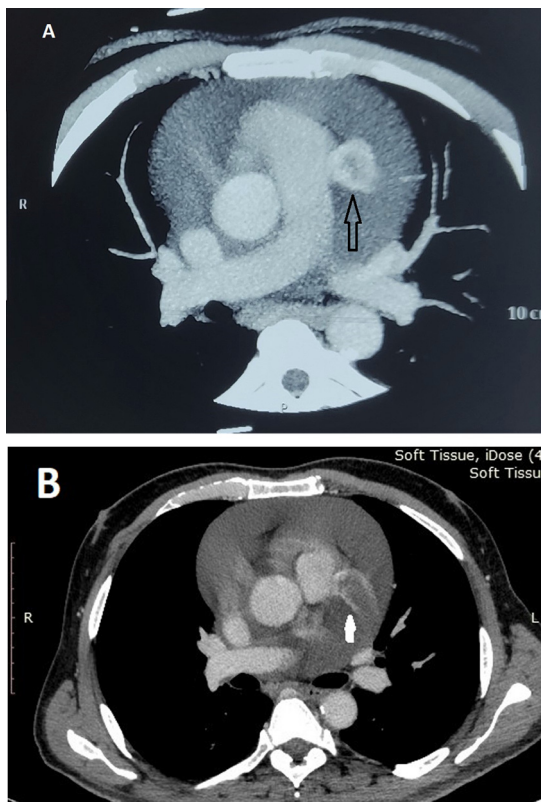


Fig. 3 – (A) contrast CT of chest showing an outpouching from pulmonary artery with contrast in it (arrow). (B) Pulmonary CT angiography showing a tumor with central isodensity.

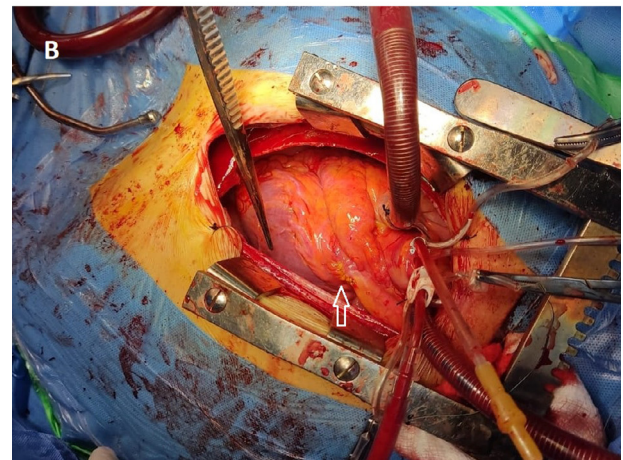
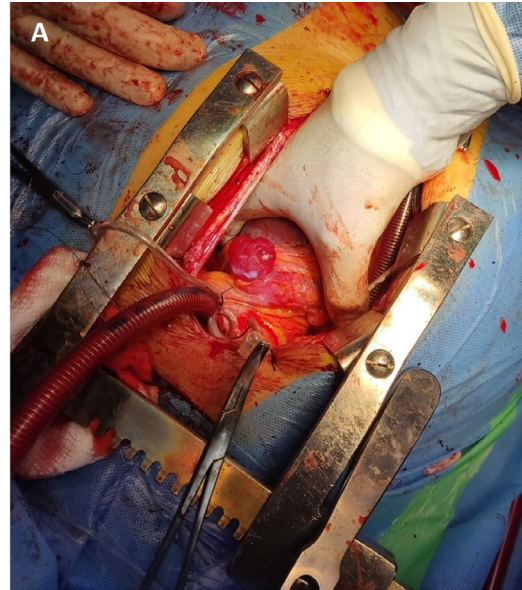


Fig. 4 – A intraoperative picture of the tumor located on the right ventricle and Pulmonary Artery B. site of the tumor after resection.

ular and pulmonary trunk contained an epicardial vascular tumor. Aortic and right atrium cannulations were performed on the patient's heart, and the patient was put on cardiopulmonary bypass since we did not know whether the invasion was intracardiac or not, and the heart was halted in diastole (Figs. 4A and B). The tumor was completely resected and sent for histopathological examination (Fig. 5). An epicardial hemangioma of mixed Capillary and Cavernous type was found during the biopsy (Fig. 6). Hematoxylin and eosin stain (H&E) was used with a magnification of (4×) We demonstrated the location of the mass in our drawing by sketching it out (Fig. 7).

The postoperative period was uneventful. The patient was discharged from the hospital after 4 days of hospitalization. Following 3 months of follow-up, neither the pericardial effusion nor the tumor was observed to recur.

The preop blood investigations included: Hemoglobin 10.7 g/dL, Platelet count of 290,000 platelets per microliter of blood,



Fig. 5 – The specimen when resected.

and WBC, 8300 WBCs per microliter. The postop investigations included: Hemoglobin 10.3 g/dL, Platelet count of 260,000 platelets per microliter of blood, and WBC, 13,800 WBCs per microliter.

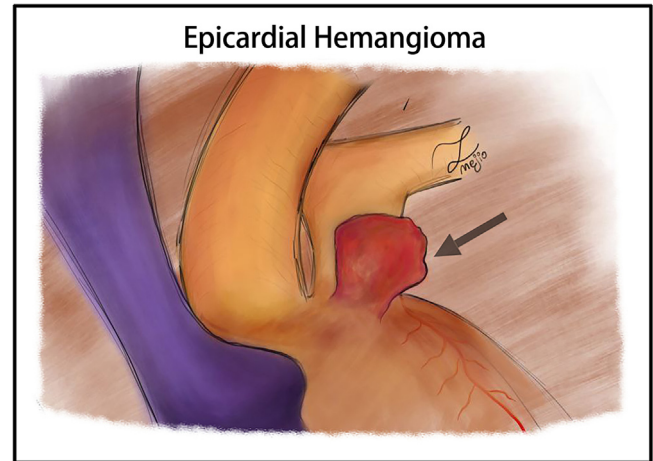


Fig. 7 – A drawing showing the location of the tumor sketched by author.

Discussion

Hemangiomas are benign, proliferative tumors characterized by an increased turnover of endothelial cells [2,3]. Cardiac hemangiomas may originate from any of the layers of the heart, including the endocardium, myocardium, and epicardium [6,7]. In addition, there have been reports of tumors originating from the pericardium. A further classification of hemangiomas is based on their histopathological characteristics, such as cavernous, capillary, arteriovenous or dysplastic types [1,6]. The cavernous type is slightly more common than other types [1]. Mixtures of these types are not uncommon.



H&E stained slides (4X)

Fig. 6 – Histopathological examination showing mixed type epicardial hemangioma.

mon [1]. The pathology of our patient's tumor revealed mixed cavernous and capillary features.

The location of such masses varies, but 39% are found in the left ventricle, left atrium, mitral valve, and aortic valve [1]. A further 44.1% of hemangiomas are found in the right heart - the most common being the right atrium [1]. Depending on the origin, the right ventricle is usually the most common site, followed by the right ventricle [5]. Most tumors are located on the epicardium rather than inside the chambers [8].

The presentation of these tumors is usually asymptomatic in the early stages unless cardiac insufficiency occurs [1]. In the majority of cases, the patient is either asymptomatic or normal on physical examination [1]. A decrease in exercise tolerance is the most common presenting symptom [1]. In benign epicardial masses, pericardial effusion is rare, whereas it occurs in benign pericardial masses - [1,2,9], especially in malignant tumors that cause bloody pericardial effusion or tamponade [1,9]. In our case, however, it was neither a bloody effusion nor a malignant tumor.

The literature rarely mentions associated diseases associated with benign epicardial hemangiomas, such as Turner syndrome, pectus excavatum, and Ebstein anomaly [1]. There have been reports of cases of MDS with splenic hemangiomas [10]. As far as we know, there are no cases in the literature in which patients with MDS are diagnosed with epicardial hemangiomas. We also could not find any correlation in the literature regarding the treatment the patient has received in the last 4 years, Danazol, and the formation of any form of hemangiomas.

It is extremely difficult to diagnose epicardial masses on the basis of clinical findings. It is still common for cases to be missed, even with the advancement of technology [1]. If contrast is used, transthoracic echocardiography should be the first modality of choice since it is a vascular tumor and will absorb the contrast rapidly [6].

Additionally, it illustrates the location and size of the tumor [8]. CT scans with contrast will also show intense enhancement. An MRI of the heart shows isointense T1-weighted images or hyperintense T2-weighted images, as compared to the myocardium [6,11]. After utilizing multiple imaging modalities in our patient, doubt remained as to the extent of the invasion.

The definitive diagnosis of such entities remains surgical resection and histopathological examination [8,11]. Because the natural history of the disease does not favor conservative treatment, surgical resection is still the treatment of choice. A high recurrence rate is observed among large hemangiomas, particularly when partial resection has been performed [8]. Even asymptomatic patients should undergo surgery in case there are life-threatening complications [7,8,11].

Conclusion

A benign primary cardiac tumor is extremely rare, and hemangiomas are among the rarest. Epicardial hemangiomas rarely present with recurrent pericardial effusions, and most recurrent pericardial effusions caused by tumors are malignant. The relationship between myelodysplastic syndrome and cardiac hemangiomas has not yet been established in the lit-

erature. Despite advancements in medical technology, cardiac hemangiomas remain challenging to diagnose. For the diagnosis and treatment of such diseases, complete resection remains the best option. We recommend that clinicians be aware of the possibility of primary cardiac tumors when treating patients with recurrent pericardial effusions without a primary cardiovascular disease.

Patient consent

Written consent was provided from the patient to the Cardiac Center's legal committee and the surgeon in charge to publish this case and show photos that include investigations and intraoperative images. If the journal so requests, these are available for review.

Ethical statement

An ethical approval has been obtained from the hospital's Ethical Committee. The patient consented to participate in the study and written consent was obtained from the patient for this case report and accompanying images. This consent is available for review upon request.

Authors' contributions

Dler is Surgeon in charge and decided for surgery, also responsible for data collection. Shkar gave scientific opinion regarding the operation and reviewed the discussion of the case report. Yad wrote and finalized the manuscript and corresponding author. Erfan reviewed literature and helped with collection of data. Zryan helped with manuscript writing and sketched the figure illustrating the tumor site. Razhan reviewed the manuscript for grammar and sentence structuring. Han is the Pathologist whom diagnosed the specimen and wrote pathology report. Othman is radiologist who wrote radiology report and defined the tumors location. All authors accepted final draft.

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