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Cardiac Epithelioid Hemangioendothelioma - A Rare Malignant Primary Cardiac Tumour: A Case Report

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Abstract **Case Report**

A 35 year old gentleman presented to a medical centre with backache and palpitation for a month. He was referred to our centre for right atrium(RA) mass. He underwent open heart surgery via median sternotomy. The large tumour was raised from interatrial septum, extending into the IVC and the postero-inferior part of RA. The inferior vena cava (IVC) was fixed posteriorly due to the tumour invasion. Debulking surgery was performed. Histopathology and immunohistochemical study confirmed a malignant hemangioendothelioma. He has completed a palliative course of tomotherapy. He is treating with Tazemetostat, an inhibitor of EZH2 methyltransferase. He remains asymptomatic from cardiac aspect after 1 year of surgery.

Keywords: Primary cardiac tumour, malignant, cardiac epithelioid hemangioendothelioma, tomotherapy, targeted therapy, Tazemetostat.

INTRODUCTION

Primary cardiac tumours are rare. The incidence rate reported as low as 0.001-0.3% found in autopsies [1]. Among these, the benign tumour composed of 75% and the remaining 25% are the malignant tumours [2,3]. Of these malignant tumours, 75% are sarcoma [3]. Here we describe an interesting case of primary cardiac epithelioid hemangioendothelioma required debulking surgery, chemoradiotherapy and targeted therapy.

CASE REPORT

A 35 year old gentleman presented to a medical centre with backache and palpitation for a month. He was referred to our centre for right atrium mass. There was unremarkable findings on physical examination. Transthoracic echocardiography (ECHO) showed dilated right atrium with a mass seems arising from inferior vena cava (IVC) and attached to interatrial septum with the biggest area of 15.2cm^2 and length of 4.7cmx 4.2cm. Computed tomography of thorax showed the lobulated enhancing mass in the right atrial chamber, measuring 4.6cmx7.8cmx7.3cm (APxWIDTHxCC) which encasing the IVC (Figure 1). There was filling defect in the superior aspect of the IVC which likely representing tumour infiltration. PET-CT showed metastatic nodes in the mediastinal, pericardial and celiac region.

Surgery was performed via median sternotomy. Aorta and superior vena cava (SVC) were cannulated. IVC was not cannulated as the tumour was huge and obstructing the IVC. Aortic cross clamp applied. Antegrade cardioplegia was given at the aortic root. He was cooled to 29°C. Right atrium was opened longitudinally. A sump sucker was inserted into IVC to drain the IVC. On the inspection, the large tumour raised from the interatrial septum, extending into the IVC, the postero-inferior part of RA. The IVC was fixed posteriorly due to the tumour invasion. Deep circulatory arrest was performed for 15minutes to create bloodless field. The tumour was excised from the RA as shown in Figure 2, weight 34grams, measuring 55mm x 40mm x 40mm (LENGTH X WIDTH X HEIGHT). The right atrium was closed by using Prolene 3/0. Intraoperative transoesopheal echocardiography (TOE) before excision and after excision showed in Figure 3.

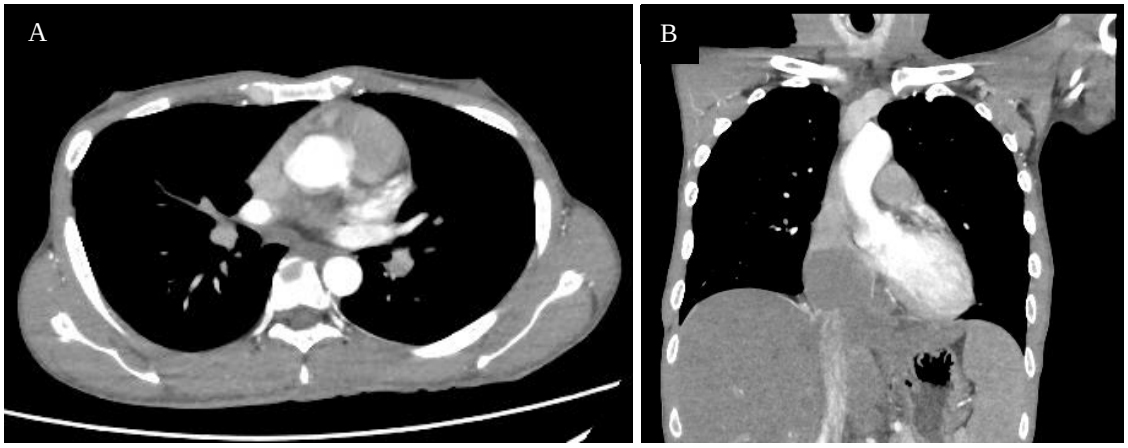

Figure 1: (A) Preoperative CT scan-axial view, (B) Preoperative CT scan-coronal view. Lobulated enhancing mass in the right atrial chamber, measuring 4.6cmx7.8cmx7.3cm(APxWIDTHxCC) which encasing the IVC.

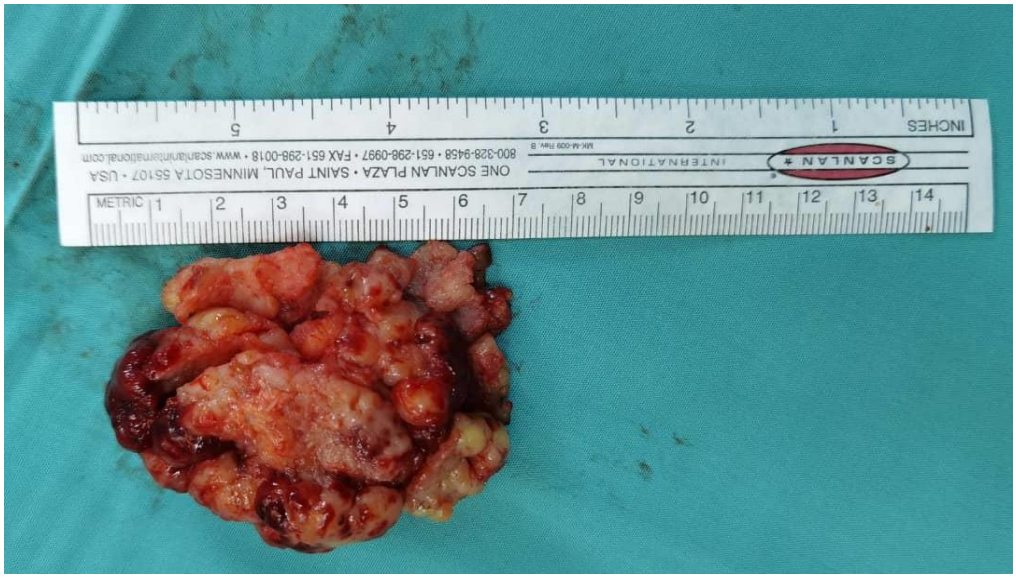

Figure 2: Right atrium mass 55x40x40mm, weights 34grams

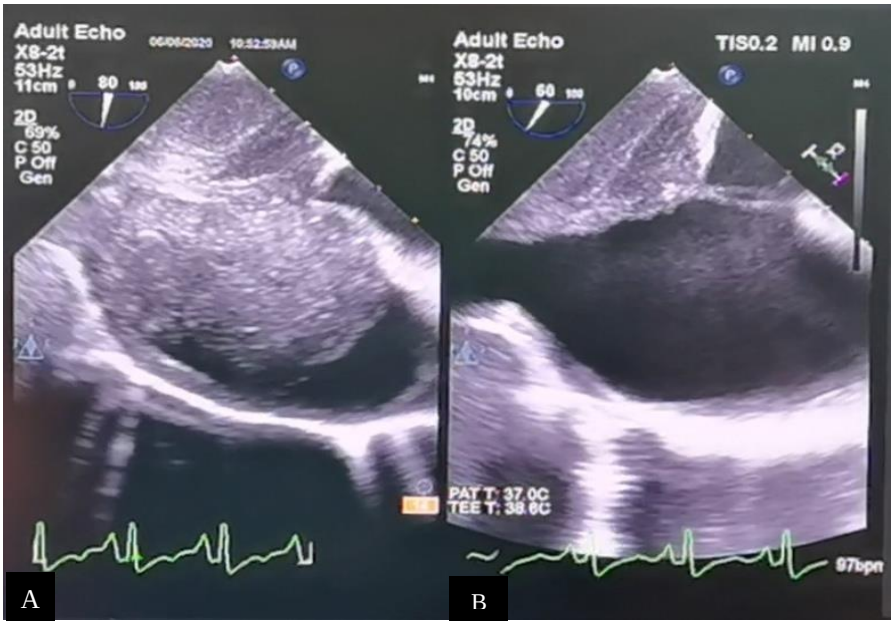

Figure 3: Intraoperative TOE images (A) Before debulking (B) After debulking.

His one month post-surgery ECHO showed good biventricular contractility, small remnant of RA mass at posterior RA wall. In histopathological study, the tumour cells appeared as malignant neoplasm with metastatic poorly differentiated carcinoma. Immunohistochemical study confirmed a malignant hemangioendothelioma.He underwent tomotherapy (TOMO) with 39Gy in 13 fractions (palliative dose), with the intention of controlling the disease. During TOMO session, the heart was avoided from the radiation (Figure 4). Post TOMO PET-CT showed reduction in the previous RA/IVC mass and the mediastinal nodes. His latest CT showed metastatic to lung, liver and spine. He is currently on Tazemetostat, an inhibitor of EZH2 methyltransferase. He remains asymptomatic from cardiac aspect.

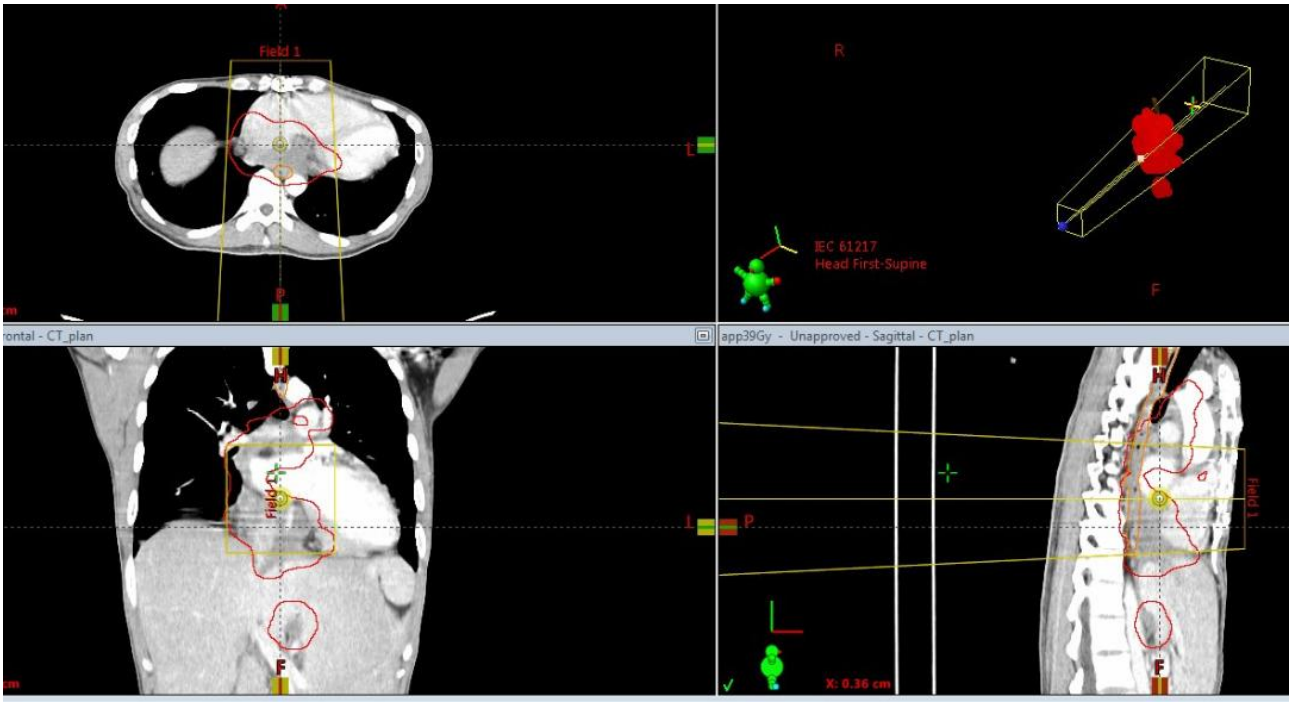

Figure 4: Tomography (TOMO) plan: red area is the planning target volume.

DISCUSSION

Epithelioid hemangioendothelioma(EHE) is a rare malignant endothelial neoplasm which usually involving soft tissue, skin, lung, liver and bone, and rarely from the heart[4]. It is extremely rare with an incidence of 1 in 1 million [5]. It was firstly described in 1982 by Weiss and Enzinger [6]. It termed as EHE as it pursues the clinical course intermediate between a hemangioma and angiosar-coma. It has 2 histological grade based on its aggressiveness: low grade (benign) and intermediate grade (malignant). The occurrence of systemic metastases reported as high as 21% [5,7]. The age prevalence of cardiac EHE 45.5±16.5 year-old[8]. According to Marzia et al study, 41% of primary cardiac hemangioendothelioma (HE) arise from right atrium with predominantly 66% epithelia type [8]. The clinical features of presentation can be varies. It usually found incidentally in asymp-tomatic patients or during cardiac assessment for symptomatic groups ,i.e: palpitation, arrhythmia, pericardiac effusion, heart failure, outflow tract obstruction or thromboembolic events [9]. The pro-gnostic factors are based on tumour's mitotic activity and its size. Size > 3.0cm indicates the worst prognosis with a 5 years survival of 59% [10]. The gold standard mainstream therapy is surgery [3,10]. In our case, we could only performed debulking surgery as the tumour has locally inva-ding into the superior aspect of IVC. There are studies shown that radiotherapy, chemotherapy, and the newer antiangiogenetic agents help to control the progression of disease. There's no proper guideline on surveillance follow up on primary cardiac tumour patient. In our centre, we review pa-tient 6 monthly and surveillance ECHO annually.

CONCLUSION

Cardiac primary EHE is extremely rare. The confirmation of the aggression type is of paramount importance. Radical surgery when it's feasible play an importance role of preventing its recurrence. Multimodality therapy may be considered in those with local or systemic metastases. Long term follow up may help us to understand more about this disease and it's prognosis.

Authors' Note

The permission was obtained to publish the case report from patient.

Ethics Approval

This is a case report. Ethic approval not applicable.

Declaration of Conflicting Interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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