Interactive CardioVascular and Thoracic Surgery (2020) 1–3
doi:10.1093/icvts/ivaa036

Cite this article as: Bislimovski D, Nikolic A, Mustafa E, Milojevic M. Symptomatic primary cardiac haemangioendothelioma during late pregnancy. Interact CardioVasc Thorac Surg 2020; doi:10.1093/icvts/ivaa036.

Symptomatic primary cardiac haemangioendothelioma during late pregnancy

Darko Bislimovski[a], Aleksandar Nikolic[a], Elvis Mustafa[a] and Milan Milojevic[a,b,*]

[a] Department of Cardiac Surgery, Acibadem Sistina Hospital, Skopje, North Macedonia
[b] Department of Cardiothoracic Surgery, Erasmus University Medical Center, Rotterdam, Netherlands

* Corresponding author. Department of Cardiothoracic Surgery, Erasmus University Medical Center, Pieter De Hoochweg 112B, 3024 BJ Rotterdam, Netherlands. Tel: +31-6-44346031; fax: +31-1-7032358; e-mail: m.milojevic@erasmusmc.nl (M. Milojevic).

Received 7 January 2020; received in revised form 29 January 2020; accepted 4 February 2020

Abstract

Primary cardiac haemangioendothelioma is an uncommon vascular neoplasm with an infiltrative growth pattern and malignant potential. The treatment of this tumour alone is challenging, but the symptomatic presentation during late pregnancy creates a complex clinical situation for both the mother and physician. To the best of our knowledge, <40 cases of primary cardiac haemangioendothelioma have been reported worldwide, but none of these were associated with pregnancy. We report a case of symptomatic primary haemangioendothelioma in the right atrium of a pregnant woman in the 36th week of gestation. A primary caesarean section, followed by complete resection of the cardiac tumour, was carried out without any complications. The mother and child were discharged home 5 days after the surgical procedure and birth, respectively. At the 2-month follow-up, positron emission tomography showed signs of hypermetabolic activity in the right atrium, without signs of tumour recurrence on the subsequent computed tomography and magnetic resonance imaging, thus a close clinical follow-up was recommended.

Keywords: Haemangioendothelioma • Late pregnancy • Right atrium

INTRODUCTION

Haemangioendothelioma is a vascular neoplasm that represents <1% of all vascular tumours, displaying a range of different biological behaviours from benign haemangiomas to the highly malignant angiosarcomas [1]. Because of its metastatic potential and infiltrative growth pattern, these tumours are classified as malignant along with the spectrum of angiosarcomas by the World Health Organization. The management of these tumours involves radical surgical resection since adjuvant chemotherapy and/or radiation have usually been found to be ineffective [2]. Primary haemangioendotheliomas are commonly found in the lung, liver and bone; however, <40 cardiac cases have been reported in the literature, of which none was associated with the pregnancy [3]. We report a case of a 34-year-old woman in the 36th week of gestation presenting with a symptomatic haemangioendothelioma in her right atrium.

CASE REPORT

A 34-year-old woman in the ninth month of pregnancy was referred to our emergency department with shortness of breath and chest pain with body position changes. The patient had a history of asthma and allergic rhinitis. The initial laboratory tests, electrocardiogram (ECG) examination, physical examination and gynaecological examination did not reveal any abnormalities. A transthoracic echocardiography was performed, revealing a large intracardiac mass in the right atrium $\sim$3.5 cm $\times$ 3.4 cm in size, which had infiltrated the interatrial septum. However, no tricuspid regurgitation or pulmonary hypertension was observed (Fig. 1A). Because of the relatively high risk for thromboembolism and/or outflow obstruction, a multidisciplinary heart team made the decision to perform an emergency caesarean section followed by a cardiac surgical procedure.

The caesarean section was performed by a gynaecological team under general anaesthesia, using transoesophageal echocardiography to facilitate perioperative monitoring and fluid management. A 2900-g female infant was born with an Apgar score of 7/8. After the caesarean section, the tumour was resected *in toto* (Fig. 1B), which was found to have infiltrated the lateral wall of the right atrium, interatrial septum and superior vena cava. After that, the right atrium and interatrial septum were reconstructed using a xenopericardial patch. The postoperative course was uneventful with excellent clinical recovery, and the mother and her baby were discharged on the fifth postoperative day.

The pathological report was suggestive for an atypical epithelioid haemangioendothelioma. Additional immunohistochemical examination demonstrated that the cells of the neoplastic proliferation expressed reactivity for CD31, CD34, WT-1, aSMA+, markers for the composite haemangioendothelioma, which is

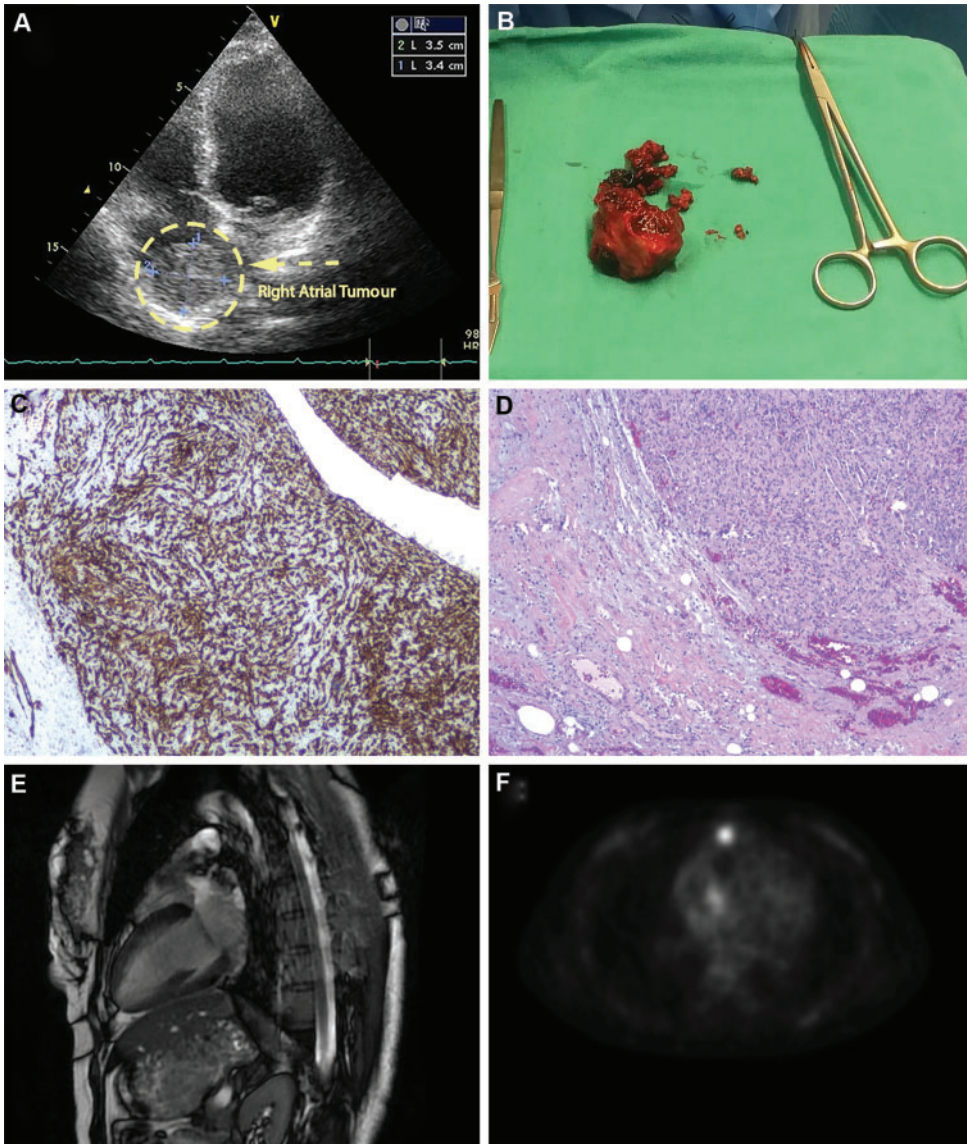

Figure 1: Transthoracic echocardiography showing a tumour (**A**) and macroscopic appearance of the resected tumour (**B**). Cardiac epithelioid haemangioendothelioma, CD31 expression and H&E, ×10 magnification (**C** and **D**). After 2 months of surgery, positron emission tomography scans revealed signs of inflammation in the area surrounding the right atrium without signs of tumour recurrence (**E** and **F**).

known as an aggressive vascular neoplasm of low-grade malignancy (Fig. 1C and D).

DISCUSSION

Primary cardiac tumours are rare, representing <5% of all cardiac tumours, and only 15% are malignant. Sarcomas are the most common malignant tumours, although other tumour types, including lymphomas, paragangliomas and extramedullary plasmacytomas, have been commonly reported in the literature. Epithelioid haemangioendotheliomas, classified as a sarcoma of vascular origin, are extremely rare, and because of its rarity, there is no standard of care. A recently published systematic review of case reports suggests that the 10-year survival of patients with primary cardiac haemangioendothelioma is generally good; however, disease recurrence is not uncommon [2]. The effectiveness of routine postoperative adjuvant therapies remains unclear and a matter of debate in this setting. Although aggressive adjuvant therapy appears to improve long-term survival after the surgical treatment of pulmonary artery and heart sarcomas [4, 5], its routine use, including the use of antiangiogenetic agents, does not have a well-established role in patients with epithelioid haemangioendothelioma. Indeed, additional adjuvant therapy may be the treatment of choice in cases where tumour margins are close or cannot be assessed, but because of limited evidence, further studies, involving laboratory experimentation, are warranted. Nevertheless, the management of pregnant patients with cardiac tumours is a particularly delicate affair since the unborn child is also at risk. Thus, multidisciplinary teamwork is needed to promptly assess gestational age, evaluate foetal viability and risks associated with a prompt surgical intervention in order to define the best treatment strategy. In the presented case, considering the minimal risk to the foetus and the mother's chances of having

severe complications before childbirth, an urgent caesarean section with subsequent surgical care was recommended by the multidisciplinary team. This patient delivered successfully, and the tumour was radically resected with no need for adjuvant therapy. The patient was discharged in a good condition and a positron emission tomography (PET) scan was performed 2 months later to avoid false-positive results from the postsurgical inflammation. The PET scan showed signs of hypermetabolic activity in the area surrounding the right atrium, without signs of the tumour recurrence based on the subsequent computed tomographic and magnetic resonance imaging scans (Fig. 1E and F). Because of its malignant potential, an extended period of close clinical follow-up was warranted.

CONCLUSION

Any cardiac disease in pregnancy is associated with a substantial risk for maternal and foetal complications. Although primary cardiac tumours are rare, when it does occur, and the foetus is viable, caesarean delivery followed by immediate open surgical repair should be the treatment of choice. A prompt multidisciplinary team approach is crucial in the successful evaluation and management of these patients.

ACKNOWLEDGEMENTS

The authors are indebted to Dzengis Jashar, pathologist at the Acibadem Sistina Hospital.

Conflict of interest: All authors report no conflict of interest.

REFERENCES

[1] Requena L, Kutzner H. Hemangioendothelioma. Semin Diagn Pathol 2013;30:29–44.
[2] Cottini M, Lappa A, Donfrancesco S. Primary cardiac hemangioendothelioma in the adult and cardiac involvement in patient with non-cardiac hemangioendothelioma. Review of the literature. Cancer Stud Ther J 2017;2:1–6.
[3] Sardaro A, Bardoscia L, Petruzzelli MF, Portaluri M. Epithelioid hemangioendothelioma: an overview and update on a rare vascular tumour. Oncol Rev 2014;8:259.
[4] Yin K, Zhang Z, Luo R, Ji Y, Zheng D, Lin Y *et al*. Clinical features and surgical outcomes of pulmonary artery sarcoma. J Thorac Cardiovasc Surg 2018;155:1109–15.
[5] Abu Saleh WK, Ramlawi B, Shapira OM, Al Jabbari O, Ravi V, Benjamin R *et al*. Improved outcomes with the evolution of a neoadjuvant chemotherapy approach to right heart sarcoma. Ann Thorac Surg 2017;104:90–6.