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Intracardiac Papillary Endothelial Hyperplasia: A Rare Case Report

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Abstract: Intracardiac papillary endothelial hyperplasia is seen very rare in the literature. This report describes the case of a 65-year old male patient who had intracardiac neoplasm behind the right coronary aortic cusp. The lesion was treated successfully by surgical excision. According to histopathological analysis the diagnosis was papillary endothelial hyperplasia. The patient was discharged without any complication on the 4th day after the operation.
Keywords: intracardiac neoplasm, papillary endothelial hyperplasia.

INTRODUCTION

Papillary endothelial hyperplasia (PEH), is a well-defined histological diagnosis however it has been reported rarely. It is a common vascular tumor of soft tissues which was first described by Pierre Masson in 1923 [1]. The lesion is thought to be the result of reactive endothelial proliferation rather than neoplasia. It is characterized by papillary lobules of proliferating endothelial cells with an underlying fibrous stroma and often resembles angiosarcoma [2]. It may occur in any blood vessel in the body but is commonly found on the fingers, head, neck, and trunk [3]. PEH may occur either in a pure primary form, as a focal change in a pre-existing vascular lesion (haemangioma, pyogenic granuloma, or vascular malformation), and rarely in an extravascular location following organization of a haematoma .

CASE REPORT

A 65-year old male patient was admitted to our clinic with exertional dyspnea which had developed over the preceding a few weeks. On admission, his blood pressure was 130/70 mmHg and his pulse rate was 75 beats/min which was regular. On auscultation, moderate diastolic murmur was heard over the right sternal border. There were no signs of heart failure. Transthoracic and transesophageal echocardiography identified a mass 1x0.8 cm in size and located beneath the right coronary aortic cusp. The ejection fraction was 58%, subaortic gradient was 34mmHg. There were no regurgitation on neither mitral nor aortic valve. We decided that thoracoabdominal computed tomography to exclude distal embolisation from the mass. Thoracoabdominal computed tomography screening was normal. No abnormality was detected on a coronary angiogram. Due to the potential of distal embolization and mild subaortic gradient we decided on the surgical excision of the mass. The operation was performed through a J sternotomy under cardiopulmonary bypass. Intraoperative transesophageal echocardiography was performed to detect the location of the mass (Figure 1). Transverse aortotomy was performed 2 cm above the coronary arteries. The mass was attached beneath the right coronary aortic cusp which was seen easily by the help of 5 mm endoscope. The lesion was treated successfully by total surgical excision. Cross clamp time was 30 minutes and cardiopulmonary bypass time was 40 minutes. The patient had an uneventful postoperative period and was discharged on the 4th day after the operation. Macroscopic assessment showed a 1 x 0.8cm ovoid mass of tissue (Figure 2). Haemotoxylin and eosin stained sections demonstrated a prominent papillary network of endothelial cells covering a loose stromal core (Figure 3). The histopathological diagnosis was papillary endothelial hyperplasia.

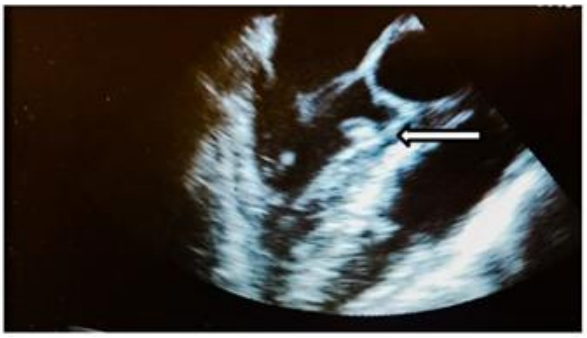

Fig-1: Intraoperative transoesophageal echocardiogram showing mass beneath the right aortic cusp.

Fig-2: Macroscopic assessment showed a 1X0.8cm ovoid mass of tissue.

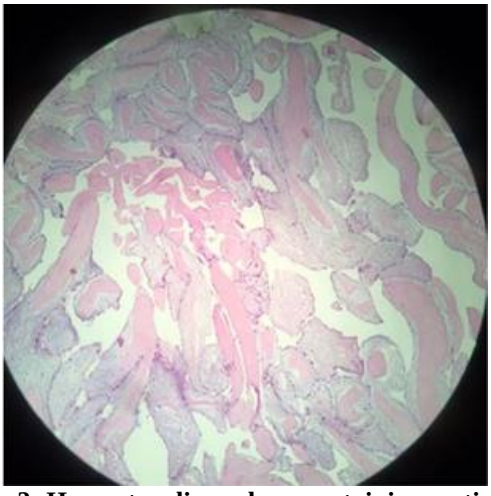

Fig-3: Haemotoxylin and eoson staining sections revealed endothelial-coverd, papillary formations.

DISCUSSION

Primary cardiac tumors are rare, and primary cardiac valve tumors are extremely rare. The reported incidence of primary cardiac valve tumors was less than 10 % of all primary cardiac tumors [4]. Myxoma was the most common benign tumor and sarcoma was the most common primary malignant tumor. The most common primary sites of cardiac metastasis were, in order of decreasing frequency, the lung and pleura, lymphoma, breast and esophagus [5]. Characteristic symptoms of intracardiac tumors include: dyspnea, chest pain, cough, hemoptysis, syncope and right ventricular failure. Echocardiography is the procedure of choice for detecting intracardiac tumors, including the more common metastatic as well as primary tumors. Papillary endothelial hyperplasia was regarded as a benign tumor of endothelial cells and can be confused with a low grade angiosarcoma [6]. Papillary endothelial hyperplasia may occur either in a pure primary form, as a focal change in a pre-existing vascular lesion (haemangioma, pyogenic granuloma, or vascular malformation), and rarely in an extravascular location following organization of a haematoma. It is a reactive proliferation of endothelial cells rather than a neoplastic growth and occurs in a variety of locations but its intracardiac occurrence is extremely rare and it has not been reported beneath the right coronary aortic cusp.

Total surgical excision is indicated for the floating mass in the left ventricular cavity because it may induce ventricular arrhythmias, sudden death, or systemic dissemination of the thrombus [7].

The lesion in our patient is most likely a pure form papillary endothelial hyperplasia beneath the right coronary aortic cusp which has not been reported so far. The lesion had neither been reported on preoperative transthoracic echocardiography nor on intraoperative transesophageal echocardiography.

CONCLUSION

This report describes a case of papillary endothelial hyperplasia beneath the right coronary aortic cusp being successfully treated by surgery. In our opinion; serious complications such as systemic dissemination, arrhythmias or sudden death can be prevented by complete surgical resection of the mass.

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