Surgical treatment of a giant cystic tumor of the atrioventricular nodal region

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Received 3 September 2008; received in revised form 14 December 2008; accepted 15 December 2008

Abstract

Cystic tumor of the atrioventricular nodal region is a rare primary cardiac tumor that can cause heart blockage and sudden death. Antemortem diagnosis and successful excision of the atrioventricular nodal region are extremely rare. A 41-year-old woman who presented with dyspnea and palpitations is reported. Electrocardiography revealed third-degree atrioventricular block. Echocardiography showed a right atrial cystic mass attached to the interatrial septum. The patient underwent surgical excision of the mass. Histopathological findings were of a cystic tumor of the atrioventricular nodal region. Placement of a permanent pacemaker was required for complete heart blockage. A two-year follow-up has revealed no sign of recurrence. This is the first case to be reported in China.

Keywords: Atrioventricular block; Cystic tumor; Excision

1. Introduction

There are a variety of lesions that can arise from the right atrium and the interatrial septum and involve the conduction system. One of them is a cystic tumor of the atrioventricular nodal region which is different from mesothelioma – the other similarly type of tumor of the atrioventricular [1].

Cystic tumor of the atrioventricular nodal region is a rare primary cardiac tumor. It can cause various degrees of heart blockage, and is the smallest tumor capable of causing sudden death [2–4]. Although they have been described in the literature, most of them were diagnosed postmortem. Antemortem diagnosis and successful excision of this type of tumor are extremely rare. Up to now, only four authenticated cases for surgical treatment have been reported. Some cases have been incidentally diagnosed at necropsy, suggesting that they can remain clinically silent [3]. Here we report a case of cystic tumor of the atrioventricular nodal region in which the tumor was detected preoperatively and successfully excised.

2. Case report

A 41-year-old woman who presented with dyspnea on effort and palpitations visited her primary care physician. On examination her pulse was 48 beats/min and blood pressure 130/70 mmHg. A resting electrocardiogram (ECG) revealed sinus rhythm and third-degree atrioventricular block, AV junctonal escape beats, premature ventricular contraction. Echocardiography revealed a right atrial cystic lesion with a broad connection with the interatrial septum (Fig. 1a).

She underwent surgery to resect the tumor under a median sternotomy and standard cardiopulmonary bypass with warm blood cardioplegia. Bialtrial transeptal approach revealed a cystic mass, 3 cm in its greatest dimension, lying within the interatrial septum and in the area of the triangle of Koch. The wall of the coronary sinus formed the lateral wall (Fig. 1b). The cyst was incised and within it there were about five smaller cysts distributed like honeycomb appearance (Fig. 1c). Approximately 3–5 ml of yellow caseous material was found within it. Rapid cytodiagnosis was done and revealed neither malignant cells nor bacteria in the fluid. After carefully checking, the wall of the cyst was completely resected. Complete atrioventricular block persisted after surgery and a permanent pacemaker was inserted 14 days later.

Histopathological examination revealed that the cyst wall was composed of fibrous connective tissue covered by a layer of squamous epithelium with partial cornification. Within the fibrous tissue were smaller cysts lined by a similar epithelium (Fig. 1d). Focally, the cyst lining cells were stratified.

The patient’s recovery was uneventful and her symptoms completely disappeared. A two-year follow-up has revealed no evidence of recurrence of the tumor.

3. Discussion

Cystic tumor of the atrioventricular nodal region was first described in 1911 [5]. It is a rare primary cardiac tumor located in the region of the atrioventricular node. Accord-
was reported in 2000 by Paniagua et al. [8]. In their case, the cystic tumor of the atrioventricular nodal region was approximately 30 mm in diameter detected preoperatively by echocardiography and MRI and was partially excised. Postoperatively the recovery was uneventful, she developed atrial flutter and various degrees of AV block with a slow ventricular rate that required the insertion of a permanent pacemaker. Kaminishi et al. recently reported the successful prevention of heart blockage by leaving the cyst wall attached to the base of the interatrial septum [9]. There was no sign of residual mass or recurrence 12 months after surgery. Saito et al. reported successful excision of a cystic tumor of the atrioventricular nodal region in 2005. Postoperation immunohistochemical staining showed that the cells of the cyst expressed carcinoembryonic antigen and epithelial membrane antigen, suggesting that they were of endodermal origin [10].

It is still controversial whether the cyst should be resected completely from the base of the interatrial septum. However, because one complication of this tumor is sudden death as a result of ventricular tachycardia or ventricular fibrillation [3, 4, 6], we believe that complete resection is essential, even if subsequent pacemaker implant is required.

Acknowledgments

This work was supported by Natural Science Foundation of China No. 30571838. We would like to thank Dr Zhang Kailun for critical review of the original manuscript.

References