Case report

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A case of right ventricular bronchogenic cyst with the clinical presentation of cerebral infarction and pulmonary embolism

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Abstract

A congenital malformation of the bronchial tree, known as bronchial cyst, is mostly found in the mediastinum or lung. Intracardiac bronchogenic cysts are quite rare and only two cases of right ventricular bronchogenic cyst have so far been reported. We herein report a case in which a cyst developed in the right ventricle of a 65-year-old Japanese male who presented with aphasia due to cerebral infarction and pulmonary embolism. The cyst was successfully removed and the patient successfully recovered following surgery.

Keywords: Bronchogenic cyst, malformation, right ventricle, patent foramen ovale, case report

Introduction

A bronchogenic cyst is a congenital malformation of the bronchial tree [1]. The cyst usually developed in the mediastinum or lung [2,3]. An intracardiac bronchogenic cyst is therefore an extremely rare malformation [4-7] and only two cases of right ventricular bronchogenic cyst have been reported [5,6]. We recently examined right ventricular bronchogenic cyst in a 65-year-old Japanese male who presented at our hospital with aphasia caused by cerebral infarction and pulmonary embolism. The cystic mass was surgically removed. We herein describe the results of the histopathologically and immunohistochemically analyses of the surgically-removed cyst.

Case presentation

A 65-year-old Japanese male with a history of diabetes was referred to our hospital due to aphasia. A neurological examination revealed aphasia and a mild disturbance of consciousness. Data from other physiological examinations, including those for blood pressure, pulse rate, respiratory rate, and temperature were within the normal limits. Auscultatory findings were normal for the lung and heart. The abdomen was soft and flat and no tenderness was observed. There was

no peripheral edema.

A large, high intensity lesion in the left middle cerebral arterial territory was detected on diffusion weighted images during brain magnetic resonance imaging (MRI) (Figure 1). Transthoracic echocardiography revealed a mass in the right ventricle and small left ventricle due to right ventricular enlargement. Transesophageal echocardiography showed no shunt or thrombus in the left atrium and ventricle. A contrast enhanced computed tomography (CT) scan revealed a cystic mass in the right ventricle (Figure 2) and a thrombus in the right pulmonary arteries (Figure 3). Cardiac MRI detected a cystic mass arising from the interventricular septum in the right ventricle. It had an intermediate and high signal intensity on T1 and T2 weighted spin-echo images, respectively (Figures 4a and 4b).

Based on these findings, the patient was diagnosed with cerebral infarction and pulmonary embolism. The right ventricular mass was considered to be a cardiac myxoma. We could not determine whether the right ventricular mass was related to the cerebral infarction because an atrial septal defect and/or a ventricular septal defect shunt were not radiographically observed in the heart.

Soon after the diagnosis, the patient received cere-bro-protection

Figure 1. Magnetic resonance images showing a left cerebral infarction (arrow).

Figure 2. A contrast enhanced computed tomography scan showing a cystic mass (arrow) in the right ventricular.

and anti-coagulant therapies. One month later, surgical excision of the cardiac mass under extracorporeal circulation was performed. The right atrium was opened and a round mass suggestive of a cystic lesion (4 cm in diameter) arising from the interventricular septum was detected in the right ventricle.

A 5 mm patent foramen ovale was incidentally found. The cystic lesion surrounded by fibrin materials like thrombi protruded from the lumen of the right ventricle. It was successfully resected, and the patent foramen ovale was closed.

Figure 3. A contrast enhanced computed tomography scan showing a thrombus (arrow) in the right pulmonary artery.

Histopathologically, the wall of the cystic lesion was lined with ciliated columnar epithelia. Bronchial glands were present under the epithelia, suggesting this was a bronchogenic cyst (**Figures 5 and 6a**). Smooth muscle and fibrous tissue with severe hyalinization and calcification were present in the cyst wall. To confirm our diagnosis of bronchogenic cyst, immunohistochemical analyses using antibodies against thyroid transcription factor (TTF)-1, CK7, Ber-Ep4, calretinin, MUC1, MUC2, MUC5AC, MUC6, synaptophysin and villin were performed. The results indicated the columnar epithelium lining the cyst wall was positive for TTF-1 (**Figure 6b**) and CK7 (**Figure 6c**), but negative for CK20 (**Figure 6d**), MUC1 (**Figure 7a**), MUC2 (**Figure 7b**), MUC5AC (**Figure 7c**), MUC6 (**Figure 7d**), Ber-Ep4 (**Figure 7e**), calretinin (**Figure 7f**), synaptophysin (**Figure 7g**) and villin (**Figure 7h**).

The patient recovered well from surgery and was discharged 16 days post-operation. As of the writing this manuscript, he remains asymptomatic.

Discussion and conclusion

We herein presented a rare case of a bronchogenic cyst in a Japanese male. Bronchogenic cysts mostly develop in the mediastinum and lung [2,3]; however, intracardiac bronchogenic cysts are extremely rare [4-7]. To the best of our knowledge, there are only two other reports published in English on right ventricular bronchogenic cyst [5,6].

The location of the bronchogenic cyst can be explained by embryogenesis [1]. Bronchogenic cyst arises from abnormal buds from the primitive esophagus and tracheobronchial tree, which do not extend to the site where alveolar differentiation occurs. Cardiac primordial exist in a place very near to the foregut or primitive tracheobronchial tree. At this time, abnormal budding of the tracheobronchial tree may migrate

Figure 4. Cardiac magnetic resonance images showing a cystic mass (arrow) arising from the interventricular septum in the right ventricle. The cystic mass has an intermediate signal intensity on a T1 weighted spin-echo image (a) and a high signal intensity on a T2 weighted spin-echo image (b).

Figure 5. A histopathological analysis of the cystic mass. The cyst is lined by ciliated columnar cells and glands are present in the cyst wall. Hematoxylin and eosin staining, bar=200 µm.

Figure 6. Histopathological (a) and immunohistochemical (b, c and d) analyses of the cystic mass. Some nuclei of ciliated columnar epithelium (a) are positive for TTF-1 (b). The cytoplasm is strongly positive for CK7 (c), but negative for CK20 (d). Hematoxylin and eosin staining (a) and immunohistochemical analyses (b-d), bars=(a, c) 200 µm; and (b, d) 100 µm.

to myocardium [4].

Clinical presentations of cardiac bronchogenic cyst are variable, ranging from asymptomatic patients to those with dyspnea, palpitations, or pre-syncope [3,8]. In the present case, fibrinous thrombi attached to the right ventricular cyst might have caused pulmonary embolism, resulting in pulmonary hypertension. Additionally, thrombi of the right ventricle migrated past the patent foramen ovale and caused cerebral infarction. Associated lesions reported in previous cases of intracardiac bronchogenic cyst include atrial septal defect [9,10], but not patent foramen ovale.

Most intracardiac bronchogenic cysts have been found by transthoracic echocardiography [7]. Echocardiography is able to provide accurate determinations of size, location, point of attachment, mobility, and hemodynamic relevance. A diagnosis is made by contrast-enhanced chest CT and cardiac MRI, which provides a non-invasive method of obtaining 3-dimensional images of masses involving the cardiac chambers and pericardium. As observed in our case, a bronchogenic cyst has an intermediate and high signal intensity on T1 and T2 weighted spin-echo images, respectively [10].

Bronchogenic cysts with an average diameter of 3 to 4

Figure 7. Immunohistochemical analyses of the cystic mass. Ciliated columnar epithelium is negative for MUC1 (a), MUC2 (b), MUC5AC (c), MUC6 (d), Ber-Ep4 (e), calretinin (f), synaptophysin (g) and villin (h). Immunohistochemical analyses, bars=200 μm.

cm contain clear or gelatinous fluid. Microscopically, they usually are lined with ciliated columnar epithelia with focal or extensive squamous metaplasia and/or extreme attenuation of this lining can occur. The wall may contain hyaline cartilage, smooth muscle, bronchial glands, and nerve trunks. In our bronchogenic cyst, there were ciliated columnar epithelia without squamous metaplasia. We did not observe any other tissues or any cartilage, or nerves. According to immunohistochemical analyses we determined the ciliated columnar cells to be bronchial epithelia positive for TTF-1 and CK7, and negative for CK20 [11].

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

Authors' contributions	KH	NW	KH2	TN	EM	TT
Research concept and design	✓	✓	✓	--	--	✓
Collection and/or assembly of data	✓	✓	✓	✓	✓	✓
Immunohistochemistry	--	✓	✓	--	--	--
Data analysis and interpretation	✓	✓	✓	✓	✓	✓
Writing the article	✓	✓	--	--	--	✓
Critical revision of the article	✓	--	--	--	--	✓
Final approval of article	✓	✓	✓	✓	✓	✓

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