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Review article

Pulmonary valvular and pulmonary arterial myxomas

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ABSTRACT

Background: Cardiac myxomas are rarely arising from the pulmonary valve or pulmonary artery.

Materials and methods: An extensive literature search generated 22 articles with 22 patients, which were taken as the material of this comprehensive review.

Results: Most patients manifested as circulatory system symptoms. A heart murmur was noted in 17 (94.4%) patients. Pulmonary valve and pulmonary artery myxomas were misdiagnosed in 4 (22.2%) patients. The myxomas were misdiagnosed in 5 (22.7%) patients. They were misdiagnosed as pulmonary valve stenosis in 2 (40%), as pulmonary artery embolism in 2 (40%), and as pulmonary valve vegetation or cardiac tumor in 1 (20%) patient. A moving mass on echo or a filling defect on computed tomography can be helpful in the establishment of the diagnosis.

Conclusions: An early surgical treatment upon diagnosis is made due to the potentials of hemodynamic disturbances and predilection of embolization. Most patients have a good prognosis following surgical treatment. The mortality rate of this patient setting can be as high as 35.7%.

Contents

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Introduction

Cardiac myxoma is the most common primary cardiac neoplasm accounting for 75% of the cases [1]. It can occur in any cardiac chambers, typically in the left atrium [2]. Right ventricular myxoma is less common [3]. Myxomas rarely arise from the pulmonary valve or pulmonary artery. The myxomas located near the pulmonary valve may influence the opening and closing of the valve leading to valvular stenosis and (or) insufficiency [3]. They may also cause misdiagnosis and unnecessary investigations and treatments. Because of the rarity and particularity of this lesion, there is not enough information on pulmonary valve myxomas to establish guidelines for management. In this article, the clinical features of the pulmonary valve and pulmonary artery myxomas are described.

Materials and methods

A comprehensive retrieval was made in the MEDLINE and China Biology Medicine Disc (CBMdisc) databases, Highwire Press, Google Scholar, Yahoo! and ''Baidu'' search engines, LILACS and J-stage for pertinent literature published between January 1950 and June 2016. The search terms were ''pulmonary artery'', ''pulmonary valve'', ''pulmonary valve annulus'', ''pulmonary valve commissure'' and ''myxoma''. Myxoma inside the pulmonary artery or pulmonary valve orifice but arising from other sites, and other types of tumors of the pulmonary valve or pulmonary artery were excluded.

Results

A careful collection of full texts of pulmonary valve or pulmonary artery myxoma resulted in 22 articles in the first four databases and search engines [3–24]; whereas no related article was generated in the latter two databases. All of these articles were referring to a single patient. Of the 22 patients, there were 13 (59.1%) males and 9 (40.9%) females with a male-to-female ratio of 1.4:1. Patients' ages were in a normal distribution (Fig. 1) with a mean of 37.0 ± 24.0 (range, 0.17–76; median, 41.5) years ($n = 22$). The male patients were much younger than females (27.6 ± 25.8 years vs. 47.0 ± 17.0 years, $p = 0.037$). The duration of the symptoms ranged from acute onset [20] to several years [14], with a mean of 12.6 ± 19.8 (range, 0.17–60; median, 3.3) months ($n = 10$) [3,6,7,9,10,12,13,16,21,22].

Of the 16 symptomatic patients, circulatory symptoms prevailed (Table 1). Four (18.2%) patients had fever, and one of them was fever of unknown origin [21].

A heart murmur was noted in 17 (77.2%) patients: a systolic murmur in 15 (88.2%), a diastolic murmur in 4 (23.5%), and a continuous murmur in 1 (5.9%) patient [16]. An abnormal heart sound was audible in 7 (31.8%) patients, including an accentuated P_2 in 3 (42.9%) patients [9,12,13], a loud A_2 in 1 (14.3%) patient [6], an accentuated S_3 (at the left sternal boarder) in 1 (14.3%) patient [7] and a P_2 split in 1 (14.3%) patient [10], and a loud and split P_2 in 1 (14.3%) patient [14], respectively. Hepatosplenomegaly was noted in 2 patients [16,21] and hepatomegaly in 2 patients [7,13].

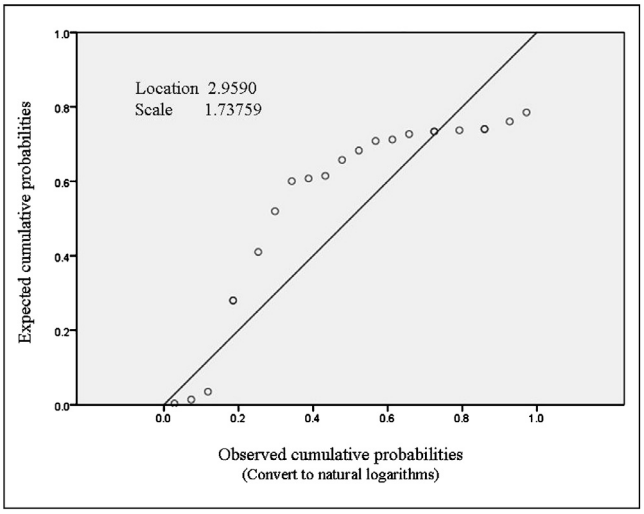

Fig. 1 – A normal distribution of patients' ages.

The associated disorders included patent ductus arteriosus ($n = 1$) [16], pulmonary valve insufficiency and stenosis ($n = 3$) [3,4,18], hypoxia ($n = 2$) [7,9], pulmonary artery or valve obstruction ($n = 2$) [10,24], pulmonary artery dilation ($n = 2$) [16,20], pulmonary embolism ($n = 1$) [9,13,20], pulmonary artery hypertension ($n = 1$) [13], hypertension and dyslipidemia ($n = 1$) [15], and complete heart block ($n = 1$) [9], myocardial bridging ($n = 1$) [14], secondary polycethemia ($n = 2$) [6,13], and multiple emboli with kidney infarctions ($n = 1$) [6].

Cytologic and microbiologic studies were done in 2 patients: gram positive cocci along with positive blood culture of *abiotrophia streptococci* was found in a resected myxoma tissue in one patient [22], and blood culture was negative in another [21].

The eletrocardiographic results were reported in 13 (59.1%) patients: normal sinus rhythm in 2 (15.4%) [3,5], atypical atrioventricular rhythm without evidence of myocardial

Table 1 – 50 clinical symptoms of 22 patients with pulmonary valve or pulmonary artery myxoma.

Symptom	*n* (%)	Reference
Dyspnea	10 (20)	[5,6,9,10,13,15,16,18,20,23]
Chest distress	4 (8)	[3,10,12,23]
Cough	4 (8)	[3,5,7,12]
Fatigue	4 (8)	[7,12,16,22]
Fever	4 (8)	[9,16,21,22]
Palpitation	5 (10)	[3,10,13,16,18]
Chills	2 (4)	[12,22]
Weight loss	3 (4)	[6,12,22]
Chest pain	4 (8)	[14,15,18,20]
Peripheral edema	3 (6)	[6,13,23]
Night sweats	1 (2)	[12]
Listlessness	1 (2)	[7]
Respiratory distress	1 (2)	[21]
Syncope	1 (2)	[17]
Dizziness	1 (2)	[14]
Fainting	1 (2)	[14]
Absence of radial pulse	1 (2)	[6]

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