**Review
Article**

Primary Cardiac Solitary Fibrous Tumors

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Primary cardiac solitary fibrous tumors were reviewed. They are classified as pericardial tumors. Their incidences are very rare. Only 16 cases were reported in the literature. Basically, surgical treatments are performed. Their prognoses are generally good, although malignant cases are also reported.

Keywords: primary cardiac tumors, solitary fibrous tumors, malignancy, CD34

Introduction

Solitary fibrous tumors (SFTs) originate from mesenchymal cells, and are spindle cell neoplasms. Although they arise in most cases from the pleura, extrapleural SFTs exist. Cardiac SFTs are rare, and are classified as pericardial tumors due to their origin.

Since SFTs mostly originate from pericardium, clinical features are similar to other intrapericardial tumors and are first diagnosed as tumors of unknown etiology. They may cause pericardial symptoms like pericarditis, tamponade, or congestive heart failure.¹⁾ Computed tomography (CT) and magnetic resonance imaging (MRI) delineate the tumors, but it is after surgical treatment that the tumors are diagnosed as SFTs.

Classification

In 1996, the Armed Forces Institute of Pathology (AFIP) classified SFT of the pericardium within benign tumors of fibrous tissue.²⁾ In 2004, the World Health Organization (WHO) classified this tumor as one of the four types of pericardial tumors.³⁾ In 2013, Amano et al. proposed a new classification in which SFTs are classified as

one of another four types of pericardial tumors and tumor-like pericardial lesions.⁴⁾ Although cardiac SFTs are rare, these classifications postulate SFTs as one entity.

Incidence

Out of 63800 cardiovascular operations performed in Japan during 2012, cardiac tumorectomy was performed in only 628 cases.⁵⁾ Out of these, cardiac SFT reported by myself in 2013 is assumed to be the sole case.⁶⁾

Many kinds of rare cardiac tumors are reported annually in medical journals.^{7,8)} Cardiac SFT is still a tumor which is reported just by finding its existence.

Cases Reported

There are only 16 cases of primary cardiac SFTs reported in the English literature, to the best of my knowledge (**Table 1**).^{2,6,9–22)} Most of them are adult cases with no gender specificity. Sizes of the tumors are relatively large indicating surgical treatment performed after being symptomatic.

The locations of the tumors vary. The origins include all the four cardiac chambers, aortic arch, and pulmonary artery. Since these tumors originate from the pericardium, the cases mentioning no specific origins are described pericardium as their locations in this table.

Surgical Treatment

Basically the treatments of these tumors are surgical. The cases which are assumed to tolerate surgical interventions are operated on. The symptomatic cases are operated to relieve their symptoms, and non-symptomatic cases to elucidate their tumor types.

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Table 1 Primary cardiac solitary fibrous tumors reported in the literature

Case no.	First author	Year	Age	Sex	Diameter (cm)	Location	Malignancy	Prognosis	Reference
1	Roggli VL	1987	NA	NA	NA	Pericardium	NA	NA	[9]
2	el-Naggar AK	1989	56	F	13.0	Pericardium	Benign	Recurrence, alive	[10]
3	Bortolotti U	1992	60	M	14	AA, PT	Benign	Alive, well	[11,12]
4	Segawa D	1995	50	F	12.5	RV	NA	Alive, well	[13]
5, 6	Burke A	1996	NA	NA	10, NA	Pericardium	Benign	NA	[2]
7	Flemming P	1996	53	F	18	LV	NA	No Mx, died post Tx	[14]
8	Andreani SM	1998	60	M	20	Pericardium	Benign	No recurrence, alive	[15]
9	Odum J	2003	NA	NA	NA	NA	Benign	No recurrence	[16]
10	Cornati G	2004	30	M	12	AA, PT	Benign	No recurrence, alive	[17]
11	Bothe W	2005	39	F	5.6	RA	Benign	Alive, well	[18]
12	Crotti UA	2008	5M	M	NA	LA	Benign	Died, non-cardiac	[19]
13	Zhao XG	2012	55	M	15	RA	Malignant	Operative death	[20]
14	Taguchi S	2013	49	F	15	LV	Malignant	Alive, well	[6]
15	Bianchi G	2013	68	F	17	LV	Benign	Alive, well	[21]
16	Tamenishi A	2013	30	F	9	ltPA	Benign	Alive, well	[22]

NA: not available; AA: ascending aorta; PT: pulmonary trunk; RV: right ventricle; LV: left ventricle; Mx: metastasis; Tx: heart transplantation; RA: right atrium; 5M: five months; LA: left atrium; ltPA: left pulmonary artery; M: male; F: female

Whether cardiopulmonary bypass (CPB) is used or not, depend on the tumor location, and the hemodynamic stability during the procedure. There are seven cases that mention the usage of CPB.^{6,13,14,17–20} There are four cases with no usage of CPB.^{11,15,21,22} Other five cases are unknown.

Prognosis

Generally, the prognoses of cardiac SFTs are good. 10 of the reported cases are alive or without recurrence. Only two reports indicate the malignancy of the cases.^{6,20} Most other reported cases are benign. In the malignant cases, hypercellularity and nuclear atypia were positive. They were stained for CD34 which are features of SFTs,²⁰ although negative CD34 is a sign of high-grade malignancy.⁶

Rare malignant examples of SFTs have been reported in the pleura and mediastinum, and analogous tumors of the pericardium have probably been classified as pericardial fibrosarcomas.^{2,6} Sarcomatous component in malignant SFTs make the diagnosis of this tumor imprecise.^{23,24}

Conclusion

Primary cardiac SFTs were reviewed. Since they are rare tumors, only 16 cases have been reported in the literature.

Disclosure Statement

The author has no conflict of interest to declare.

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