Scholars Journal of Medical Case Reports

OPEN ACCESS

Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

Unusual Localization of Fibroelastoma of the Heart in Patient with Previous Chest Radiotherapy

El-Alaoui Mohamed[1*], Fuzellier Jean Francois[1], Vola Marco[1], Campisi Salvatore[1]

[1]Cardiovascular Surgery Departement, University Hospital of Saint Etienne, Avenue Albert Raimond, 42270 Saint-Priest-en-Jarez, France

DOI: 10.36347/sjmcr.2022.v10i03.009 | **Received:** 19.03.2021 | **Accepted:** 30.04.2021 | **Published:** 11.03.2022

***Corresponding author:** El-Alaoui Mohamed
Cardiovascular Surgery Departement, University Hospital of Saint Etienne, Avenue Albert Raimond, 42270 Saint-Priest-en-Jarez, France

Abstract **Case Report**

Cardiac papillary fibroelastoma is a rare, benign, slow –growing tumor of endocardium that my have a malignant propensity for live-threatening complications. The histogenesis remains controversial; an iatrogenic origin has been suggested. Localization in the left ventricular outflow tract is extremely rare**.** We describe a case of left ventricle fibroelastoma in patient with previous chest radiotherapy, seeming to support the iatrogenic hypothesis.

Keywords: Fibroelastoma, Chest radiotherapy, Hodgkin's lymphoma.

INTRODUCTION

Cardiac papillary fibroelastoma (CPFE) is a rare, benign, slow-growing tumor that may have a malignant propensity for life-threatening complications such as thromboembolism, coronary ostia occlusion or mechanical interference with valvular function [1]. CPFE can potentially affect all cardiac structures covered by endothelium but is predominantly localized on the atrioventricular or semilunar valves with a preference for the left side valves [2]. The histogenesis remains controversial. Based on microscopic and developmental features, an iatrogenic origin has been suggested [3, 4]. Localization in the left ventricular outflow tract (LVOT) is extremely rare; we describe a case of left ventricle fibroelastoma in patient with previous chest radiotherapy for Hodgkin's lymphoma and secondary malignant fibro-papillary thyroid neoplasia, seeming to support the iatrogenic hypothesis.

CASE REPORT

A 55-year-old patient was referred to our University Hospital of cardiologic check-up. He presented a 1-month-lasting dyspnea, clinical antecedents of Hodgkin's lymphoma in the youth treated by radiotherapy and secondary papillary thyroid cancer with necessity of total thyroidectomy and new neck and chest radiation ten years earlier about. Moreover, five years before, he presented a transient ischemic attack (TIA) with left facial paralysis and dysphasia but he underwent any investigation.

On admission, physical examination, laboratory tests and electrocardiogram did not reveal abnormalities. Transthoracic echocardiography detected a mobile, avascular mass originating from the left ventricle free wall (maximum length and width of 2.25 and 1.62 cm, respectively), located immediately below the aortic valve under the right coronary cusp. The left ventricular ejection fraction, aortic and mitral valves were normal. Urgent surgery was performed under cardiac circulatory bypass and cardioplegic arrest (mean times 45 and 33 min, respectively), the tumor was radically removed through a trans-aortic valve approach, by retracting and preserving the aortic valve leaflets for the exposure. Macroscopic feature was consistent with a ''sea anemone-like'' mass with ''finger-like'' offshoots. The perioperative trans-esophageal echography showed no residual mass in LVOT. The postoperative course was uneventful and the patient was discharged at the 6th postoperative day.

Histogical examination identified the mass as a papillary fibroelastoma showing a fibrous axis with characteristics finger-like projections covered by regular endothelium.

DISCUSSION

CPFE is the third most common primary cardiac tumor [1]. It is generally localized on the atrioventricular or semilunar valves, although it can originate from all endocardial surfaces [2]. CPFE represent about 10% of all cardiac tumors in autopsy series [1], conversely, it can be discovered fortuitously or because of coronary ostia dynamic obstruction, valvular function disruption, pulmonary or cerebral embolization. Nowadays the widespread use of echocardiography allows diagnosis during life with increasing frequency. The real nature of CPFE is still under debate. Scientific evidences suggest that it could be the result of exceeding abnormal response of the endocardium to various types of stimuli. Previous cardiac surgery or chest irradiations has been supposed to be linked to CPFEs development. Ngaage et al., [3] in a series of 22 iatrogenic CPFEs found previous open-heart surgery in sixteen (73%) and thoracic irradiation in six (27%) patients, respectively. This investigation supports the concept that at least some CPFEs can represent '' post-traumatic tumors''. In the same way Kurup et al., [4] described a close association between lesion's development site and previous open-heart surgery or thoracic irradiation, so that, while the location in the cardiac chambers remaining very rare, in the group of patients with previous open-heart surgery the lesions were typically localized in the left ventricular sections in close proximity with valvular or sub-valvular structures, instead the radiation-induced lesions were localized exclusively in the right chambers. Effectively, it seems to be plausible that CPFEs may represent a delayed manifestation of radiation-induced damage of the endocardium, on the other hand the sensitivity of cardiac tissue to radiation is known and long-term survivors of Hodgkin's lymphoma can experience second malignant neoplasms and cardiovascular diseases as late adverse consequences of radio-toxicity, usually from 5 to at least 35 years after chest irradiation [5]. In our case, the differential diagnosis of the mass was between primitive cardiac tumor and malignant secondary growth of fibro-papillary thyroid carcinoma. Although cardiac magnetic resonance is the first preferable option to evaluate the nature of cardiac asses, in consideration of highly mobile feature and its localization,we decide to perform an urgent cardiac operation without further investigations.

Some Authors consider CPFEs as the result of mechanical chronic repetitive low-grade trauma in the same way that age-related lamb's excrescences or chronic forms of viral endocarditis, based on evidence of presence of cytomegalovirus (CMV) remnants in the intermediate layer of lesion's tissue samples [6]. With regard to this theory, we think in our case medical antecedent of papillary thyroid cancer could have orientated reasoning on the possible role of CMV infection but the relationship between CMV infection and thyroid cancer is not well established [7].

We found in literature a very few anecdotal reports of CPFEs localized in LVOT [8] but in any case a correlation with previous chest radiotherapy has been described for this unusual location, so that, even the location in the left cardiac chambers could be consistent with previous chest irradiation, supporting strongly the iatrogenic nature of some CPFEs. The slow-growth potential of CPFE, its location in high pressure and flow chambers of the heart, the continuous offering of a '' dysfunctional'' endothelial substrate to macro or micro-thrombi aggregation could be associated with high thromboembolic risk even in case of small lesions, so that, regard to the diagnosis, a high index of suspicion is needed, since it is not uncommon that patients experience mild neurologic complications, as in our case.

Thus, we strongly recommend urgent surgical treatment of CPFEs in all cases, independently of size and location, even in asymptomatic patients, especially since a complete excision is curative and feasible through a valve sparing technique and allows providing definitive histological diagnosis of the mass.

CONCLUSION

Cardiac papillary fibroelastoma (CPFE) is a rare, benign, slow-growing tumor that may have a malignant propensity for life-threatening complications such as thromboembolism. The management for asymptomatic patients remains controversial, though surgical excision is an accepted approach in symptomatic patients to prevent further embolic events.

REFERENCES

1. Edwars FH, Hale D, Cohen A, Thompson L, Pezzella AT, Virmani R.Primary cardiac valve tumors. Ann Thorac Surg. 1991 Nov, 52(5):1127-31.
2. Gowda RM, Khan IA, Nair CK,Mehta NJ, Vasavada BC, Sacchi TJ. Cardiac papillary fibroelastoma: a comprehensive analysis of 725 cases. Am Heart J. 2003 Sep, 146(3):404-10.
3. Ngaage DL,Mullany CJ,Daly RC, Dearani JA, Edwrs WD,Tazelaar HD, McGregor CG, Orszulak TA, Puga FJ, Schaff HV, Sundt TM 3rd, Zehr KJ.Surgical treatment of cardiac papillary fibroelastoma: a single center experience with eighty-eight patients. Ann Thorac Surg. 2005 Nov,80(5): 1712-8.
4. Kurup AN, Tazelaar HD, Edwards WD, Burke AP, Virmani R, Klarich KW, Orszulak TA. Iatrogenic cardiac papillary fibroelastoma: a study of 12 cases (1999 to 2000). Hum Pathol. 2002 Dec, 33(12):1165-9.
5. Van Leuuwen FE, Ng AK. Long-term risk of second malignancy and cardiovascular disease after Hodgkin lymphoma treatment. Hematology Am Soc Hematol Educ program. 2016 Dec 2, 2016(1):323-330.

6. Grandmougin D, Fayad G, Moukassa D, Decoene C, Abolmaali K, Bodart JC, Limousin M, Warembourg H. Cardiac valve papillary fibroelastomas: clinical, histological and immunohistochemical studies and a physiopathogenic hypothesis. J Heart Valve Dis. 2000 Nov, 9(6):832-41.
7. Huang TS, Lee JJ, Cheng SP. No evidence of association between human cytomegalovirus infection and papillary thyroid cancer. World J Surg Oncol. 2014 Feb 21,12:41.
8. Vivacqua A, Shafii A, Kalahasti V, Tan C, Gonzalez-stawinski G. images in cardiology. Ventricular outflow tract papillary fibroelastoma presenting with non-ST-segment elevation myocardial infarction. J Am Coll Cardiol. 2010 Jun 8, 55(23):2607.