

Case Report

Left atrial myxoma with papillary fibroelastoma-like features

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Abstract: Although rare, papillary fibroelastoma (PFE) of the heart valves and atrial myxoma represent the two most common cardiac tumors. Coexistence of these two lesions has been documented in rare case reports. We describe the case of a 77-year-old man who presented with slightly progressive chest pain associated with dyspnea, fatigue and edema of the lungs. Transthoracic echocardiography detected a left atrial mass that has been successfully excised. Histopathological examination showed a neoplasm combining features of both atrial myxoma and PFE. However, close evaluation of the latter showed microscopic foci of myxomatous tissue within papillary cores, indicating that the PFE-like component has developed around preexisting myxomatous tissue that served as a nidus for papillary fronds, probably by a process of fibrinous microthrombosis, organization and endothelialisation. This unusual case may shed light on the pathogenesis of the PFE pattern.

Keywords: Papillary fibroelastoma, myxoma, atrium, calretinin

Introduction

Primary tumors of the heart are rare entities among cardiac diseases. Their frequency in autopsy series was 0.0017–0.28% [1]. About 75% of primary cardiac tumors are benign; atrial myxomas being the most common [1]. The remainder was predominantly benign hemangiomas [1]. As a consequence of the major achievements made by cardiac surgery in the last decades, cardiac tumors of different histogenetic types have been increasingly recognized and treated during the patient's life. Accordingly, several larger series of surgically resected cardiac tumors have been published in the English literature [2]. On the other hand, the incidental detection of cardiac tumors at autopsy is diminishing.

Papillary fibroelastoma (PFE) is a rare cardiac tumor that predominantly originates from the heart valves [3]. This usually small lesion shows a characteristic echocardiographic, gross, and histological appearance [4]. Grossly, PFE strikingly resemble the appearance of a sea anemone upon immersion in water [4]. Histologically,

PFE is composed of thin branching papillary fronds covered by a thin layer of flat endothelium followed by a subendothelial mucopolysaccharide material that contains a central core of granular elastotic tissue [3].

Coexistence of histogenetically distinct primary cardiac tumors is rare. Documented examples of this phenomenon include myxoma concomitant with PFE [5-7] and PFE co-incidental with atrioventricular node tumor [8]. Here we present the diagnostic evaluation and successful surgical resection of a left atrial myxoma that displayed a prominent PFE-like pattern mimicking two different lesions.

Case report

A 77-year-old Caucasian man with an unclear left atrial mass was referred to our hospital for further diagnostic evaluation. The mass was detected by transthoracic echocardiography performed by his general practitioner. The patient noted slightly progressive chest pain associated with dyspnea, fatigue and oedema of the lungs. At admission to hospital he presented a

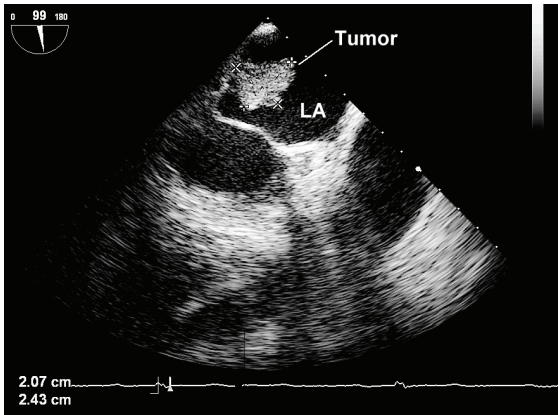


Figure 1. Transesophageal echocardiography revealed large heterogeneous, lobulated echogenic mass attached to the interatrial septum (LA, left atrium).

blood pressure of 150/70 mmHg and a heart rate of 76 beats per minute. Transthoracic and transesophageal echocardiography revealed a large mobile left atrial mass (maximum diameter: 24 x 21 mm) on the interatrial septum (**Figure 1**). The echocardiography showed no impairment of either left ventricular or right ventricular systolic function but a persistent foramen ovale was seen. The heart valves appeared normal without any signs of regurgitations or stenosis. Subsequently, selective coronary angiography was performed; there was no significant stenosis at the coronary arteries.

The patient was taken to the operating theatre where a median sternotomy was performed and cardiopulmonary bypass was installed via aortobicaaval cannulation. The large gelatinous tumor mass was successfully excised. Before leaving the hospital on day 8 after surgery, echocardiography showed a normal left and right ventricular function with an ejection fraction of 60%, well-functioning heart valves, and no pericardial effusion.

Pathological findings

The resection specimen was composed of a piece of gelatinous tissue 24 x 21 x 10 mm in maximum diameter (**Figure 2**). The surface of the tumor revealed a partial villous property. Histological examination showed a tumor composed of two closely associated components (**Figure 3A**). One component showed prominent



Figure 2. Intraoperative gross photograph showed a gelatinous tumor mass (maximal diameter of 24 x 21 mm).

papillary frond-like structures very reminiscent of PFE (**Fig. 3B**). While intermingling of both tumor components in an alternating fashion was evident (**Fig. 3B**), the papillary component was focally distinct and was almost indistinguishable from true PFE including the presence of central elastic tissue within the fronds (**Figure 3C**). The papillary structures were covered partially by flattened endothelial-like cells and occasionally by small aggregates of hyperchromatic enlarged myxoma-like cells (**Figure 3B, D**). Interestingly, close evaluation of the small papillae showed minute myxomatous foci within the papillary cores (**Figure 3D**). Microthrombi composed of fibrin and erythrocytes surrounded some of the papillary structures. The second tumor component represented a typical myxoma composed of slender or plump “myxoma cells” forming nests, communicating cords and perivascular aggregates within a prominent myxoid matrix containing degenerative elastotic material and old hemorrhages (**Figure 3E**). Immunohistochemistry showed strong expression of calretinin in the myxoma cells including those covering the papillae and intra-papillary myxomatous foci (**Figure 3E, F**). Tumor cells stained variably for CD31 and CD34. Notably, flat endothelial cells on the papillary surface did not

Combined myxoma-fibroelastoma

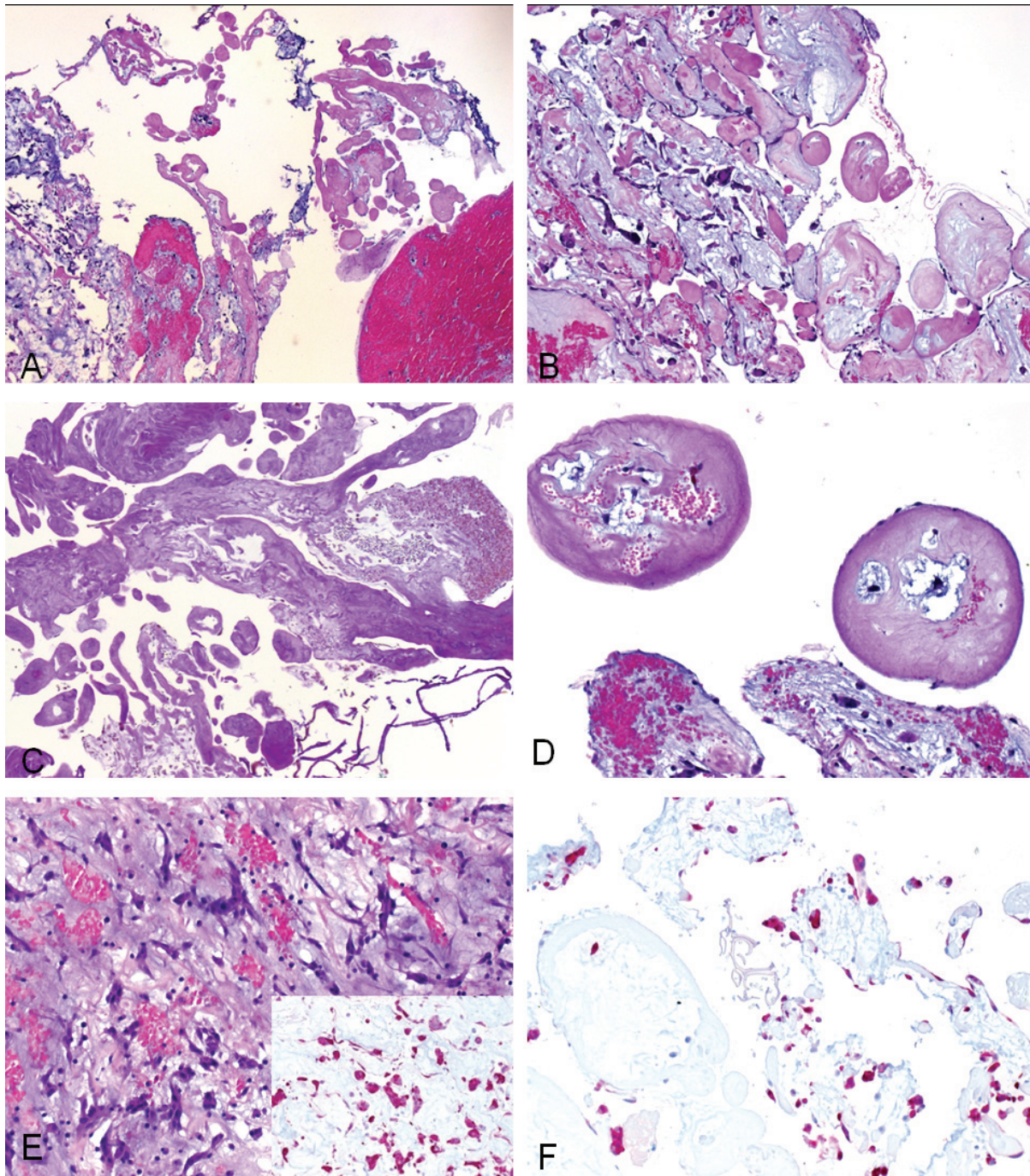


Figure 3. (A) Histological examination showed typical myxomatous tissue (lower left) adjacent to papillary frond-like structures very reminiscent of papillary fibroelastoma (upper right, note intervening thrombotic material). (B) Papillary structures alternating with hyperchromatic myxoma cells, note myxomatous cores within some papillae. (C) Elastic van Giesson stain in this area of the tumor was indistinguishable from PFE with some darkly staining granular elastic material. (D) Higher magnification of the papillae showed minute myxomatous foci (note residual bleeding and fibrin in some papillae). (E) The tumor in other areas displayed classical myxomatous appearance and expressed strongly calretinin (subimage). (F) Calretinin immunostaining highlighted myxoma cells covering the papillae and within papillary cores (note non-staining flat endothelial cells covering the papilla on the left).

stain for calretinin (**Figure 3F**).

Discussion

The histogenesis of both cardiac myxoma and PFE has been a matter of controversy. For PFE, a hamartomatous derivation [3], degenerative origin similar to the Lambi's excrescences [9], thrombotic origin and congenital as well as other theories have been suggested and discussed by different authors [10]. PFE represents the most common tumorous lesion of cardiac valves. Rare cases may arise from the atrial endocardium away from the valves and the valves leaflets [11]. An unusual case of multifocal (tapete) PFE was reported recently [10]. Although most PFEs are under 1 cm in diameter, rare giant examples have been reported [12]. A recent review revealed a total of 833 PFEs reported in the English literature between 1997 and 2008 reflecting a high awareness of this tumor that on occasion may give rise to serious complication, in particular cerebrovascular embolisation [10].

The observations that most PFEs arise in diseased cardiac valves or after a history of instrumentation or previous irradiation make it likely that an initial minute lesion (nidus) is necessary for further growth of the lesion. Accordingly, it is not excluded that the presence of common fibrous thrombotic material on the surface of some PFE might contribute to further growth of the lesion by a process of thrombosis, organization and endothelialisation of surface thrombi. The turbulent blood flow related to the localization of the lesion might be involved as well. Thus, a similar pathogenesis might be responsible for the peculiar PFE-like component in our current case. In our case, the papillary cores contained minute foci of myxoma cells indicating that these foci have served as a nidus for the development of further papillae, probably by a continuous process of microthrombosis, organization and endothelialisation. In line with this hypothesis, fresh thrombotic material was seen between and covering the myxomatous papillary structures in our case. Interestingly, a few papillary structures in our case showed foci of granular elastotic material similar to true PFE. However, the intermingling of the two tumor components and the presence of unequivocal myxomatous foci within the papillary cores definitely excluded the possibility of two histogenetically distinct neoplasms: myxoma and PFE.

Notably, all lesional cells in the myxomatous and the papillary component including also most of cells covering the papillae strongly expressed calretinin which is a known marker of myxoma cells [13]. Negativity of some of the covering cells for calretinin (as depicted in Fig. 3F) and expression of CD31 and CD34 in these cells indicated true endothelial cells.

In summary, we described an unusual case of left atrial myxoma that displayed a prominent PFE-like tumor component. The findings in this case are in line with a damage-thrombosis-organization-endothelialisation consequence in the pathogenesis of PFE and PFE-like lesions.

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References

- [1] Burke AP, Virmani R. Tumors of the heart and great vessels. Atlas of Tumor Pathology, third series, fascicle 16. Armed Forces Institute of Pathology, Washington, DC, 1996.
- [2] Blondeau P. Primary cardiac tumors. French studies of 533 cases. Thorac Cardiovasc Surg 1990;38:192-195.
- [3] Raeburn C. Papillary fibro-elastic hamartomas of the heart valves. J Pathol Bact 1953;65:371-373.
- [4] Strecker T, Agaimy A, Marwan M, Zielezinski T. Papillary fibroelastoma of the aortic valve: appearance in echocardiography, computed tomography, and histopathology. J Heart Valve Dis 2010;19:812.
- [5] Prifti E, Bonacchi M, Salica A. Mitral valve myxoma concomitant with papillary fibroelastoma. Ann Thorac Surg 2000;70:335-6.
- [6] Jallad N, Parikh R, Daoko J, Albareqdar E, Al-Dehneh A, Goldstein J, Shamon F, Connolly MW. Concurrent primary cardiac tumors of differing histology and origin: case report with literature review. Tex Heart Inst J 2009;36:591-3.
- [7] Matsushita A, Manabe S, Tabata M, Fukui T, Shimokawa T, Amano J, Takashi S. Heterogeneous double cardiac tumor: myxoma concomitant with papillary fibroelastoma. J Card Surg 2010;25:35-7.
- [8] Agaimy A, Mandl L. Papilläres Fibroelastom der Aortenklappe koinzident mit zystischem Tumor des Atrioventrikularknotens. Pathologie 2000;21:250-4.
- [9] Boone SA, Campagna M, Walley VM. Lambi's excrescences and papillary fibroelastomas: Are they different? Can J Cardiol 1992;4:372-376.

Combined myxoma-fibroelastoma

- [10] Law KB, Phillips KR, Cusimano RJ, Butany J. Multifocal "tapete" papillary fibroelastoma. *J Clin Pathol* 2009;62:1066-70.
- [11] Uchida S, Obayashi N, Yamanari H, Matsubara K, Saito D, Haraoka S. Papillary fibroelastoma in the left ventricular outflow tract. *Heart Vessels* 1992;7:164-167.
- [12] Crestanello JA, Orszulak TA. Giant papillary fibroelastoma of the right atrium: an unusual presentation. *Ann Thorac Surg* 2002;74:1252-4.
- [13] Terracciano LM, Mhawech P, Suess K, D'Armiendo M, Lehmann FS, Jundt G, Moch H, Sauter G, Mihatsch MJ. Calretinin as a marker for cardiac myxoma. Diagnostic and histogenetic considerations. *Am J Clin Pathol* 2000;114:754-9.