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Case Report

Atypical Location of a Non-Valvular Papillary Fibroelastoma: the Coumadin Ridge -

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ABSTRACT

Papillary fibroelastoma, a primary cardiac tumour commonly found on the cardiac valves is less frequently found on the non-valvular myocardium. Most non-valvular papillary fibroelastomas are confined to the left ventricle and the atrial origin of this tumour is rare. We present a case of papillary fibroelastoma originating from the left atrial ridge or the Coumadin ridge (the confluence of the long axis of the left upper pulmonary vein with the roof of the left atrial appendage). There are a few sporadic reports of the origin of papillary fibroelastomas from the left atrial ridge and the majority of these have presented with cerebral embolism. These uncommon tumours have a predilection to cause embolic phenomenon, especially those arising from the left atrial ridge.

Keywords: Left atrial ridge; Papillary Fibroelastoma (PFE); Cardiac tumour

INTRODUCTION

Papillary Fibroelastoma (PFE) is a benign cardiac tumour arising from the valvular endocardium, rarely found to originate from the non-valvular endocardium. The origin of this tumour from the left atrial ridge (the confluence of the long axis of the left upper pulmonary vein with the roof of the left atrial appendage) or the Coumadin ridge, is exceptionally rare and typically these tumours have presented with embolic phenomenon.

Papillary fibroelastoma's have also been implicated as the cause of myocardial ischemia, infarction and pulmonary embolism. We report a case of papillary fibroelastoma arising from the left atrial ridge presenting with a cerebrovascular accident.

CASE REPORT

A 61 year old male was admitted to hospital following an unwitnessed fall at home. He was found incontinent of both urine and faeces, with high blood sugars and paraparesis of his lower limbs. A CT scan showed multiple cerebral infarcts related to an embolus and an MRI confirmed bilateral acute posterior cerebral artery territory infarction (cerebellar, occipital and temporal).

A Transesophageal Echocardiogram (TOE) revealed a 23x18mm large irregular mobile mass attached to tip of the Coumadin ridge (Figure 1A). A cardiac MRI and CT more accurately delineated the intracardiac mass (Figure 1B and 1C).

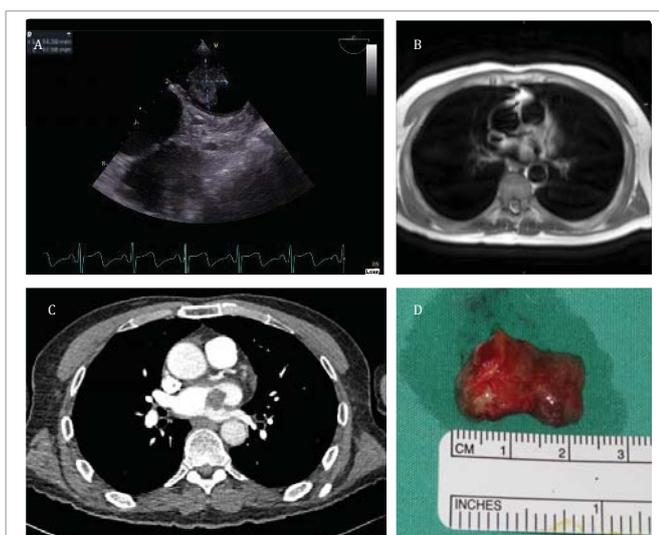


Figure 1: (A)TOE mass from the Coumadin ridge; (B) MRI - enhances the PFE in the left atrium; (C) CT showing a mass in the left atrium; (D) Photo of resected specimen

Surgery was expedited and under cardiopulmonary bypass the Left Atrium (LA) was approached through the Sondergaard's groove. The mass was found attached to the left atrial ridge between the left superior pulmonary vein and the left atrial appendage. The mass was excised along with 1cm of surrounding endocardium and myocardium necessitating primary closure. The patient made a good recovery from the surgery without any perioperative complications.

The histopathology findings were consistent with a papillary fibroelastoma.

DISCUSSION

Papillary fibroelastoma is the third most common primary cardiac tumour after myxoma and lipoma. It is the most common tumour of the valvular endocardium represented in autopsy series, accounting for 80% of the cases [1]. The etiology is unknown, however mechanical trauma associated with chronic heart disease has been postulated. Others suggest they represent a neoplasm, hamartoma or inflammatory nodule.

Cerebrovascular symptoms are the most common, with 53% of patients presenting with transient ischemic attacks or stroke [1]. The clinical spectrum varies from incidental findings to myocardial infarction, pulmonary emboli and sudden death. Papillary fibroelastoma's are friable, often with adherent thrombus, explaining their propensity to embolise [1,2].

Oda et al. [3] mentions, the speed of the blood turbulence at the LA ridge must be higher at the LA appendage or the free wall, hence PFE's originating from the LA ridge may cause embolic events more frequently.

Hirose et al. [4] reported 15 cases of fibroelastoma arising from the left atrial endocardium and interestingly 7 of these arose from the left atrial ridge, having presented with an acute stroke or history of a CVA.

Gowda et al. [2] reviewed 725 cases of PFE and found only 10 tumour's originating from the left atrium. The vicinity of the left pulmonary vein and the left atrial appendage being the most frequent location of PFE within the left atrium, with 3 cases being described on the ridge between the left atrial appendage and the left upper pulmonary vein.

Reviewing the literature has found 6 cases since 2009 (Table1). There have been no reported cases of recurrence after complete surgical removal.

Differentiating these benign lesions from those of a left atrial thrombus and urgent identification of the intracardiac mass as a PFE is vital as it represents a surgically correctable cause of an embolic phenomenon. There is general consensus that symptomatic tumour's



Table 1: Cases of PFE originating from the left atrial ridge reported since 2009.

Year	Author	Paper Title	Age	S	Symptom	Size
2010	Atalay MK	Gradual Enhancement of a Large Left Atrial Papillary Fibroelastoma on Cardiac Magnetic Resonance - The Waiting Game [5]	70	M	N/A	3 x 2.6 x 2.9
2013	Saitoh Y	Left Atrial Papillary Fibroelastoma Accidentally Diagnosed with Gastric Cancer [6]	78	F	Incidental finding	1.7 x 1.0
2013	Bashir A	Warfarin Ridge. An Unusual Location of Benign Papillary Fibroelastoma [7]	69	M	Collapse	1.8 x 1.4
2014	Waziri F	Left atrial papillary fibroelastoma as an unusual cause of myocardial infarction [8]	70	F	MI	0.9 x 1.5
2014	Hua A	Minimally invasive robotically assisted surgical resection of left atrial endocardial papillary fibroelastomas [9]	89	F	CVA	N/A
2015	Oda T	A surgical case of papillary fibroelastoma originating from the left atrial ridge [3]	49	M	CVA	0.8 x 0.8

should be surgically resected; the treatment of asymptomatic patients is debated. Surgical resection should be performed as soon as possible to avoid embolism. Asymptomatic patients should be treated surgically if the tumour is mobile, as tumour mobility is the independent predictor of death or nonfatal embolization [2].

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