

## Clinical Case Report

## Brainstem stroke caused by left atrial cardiac papillary fibroelastoma: an increasingly recognized rare cause of stroke



Luke Bonavia \*, Justin Jackson, Jennifer Pui-Yee Law, Michael Yii, Dhaval Joshi

Albury Base Hospital NSW, Epworth Eastern Hospital and Melbourne Pathology Victoria, Australia

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## ABSTRACT

Cardiac papillary fibroelastoma is a rare but increasingly recognized cause of embolic stroke that is prevalent in the older population and requires prompt surgical management. We report an unusual case of left atrial appendage cardiac fibroelastoma in a 76-year-old gentleman who presented with left internuclear ophthalmoplegia and ataxia, with corresponding diffusion-weighted imaging on magnetic resonance imaging of the brain. This case illustrates the importance of echocardiographic imaging in the workup of cardioembolic stroke in the older adult population in the acute setting.

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## 1. Case

We report a case of a 76-year-old gentleman presenting with posterior territory infarcts caused by cardiac papillary fibroelastoma (CPF) of the left atrial appendage. He presented initially with sudden severe vertigo, diplopia, and left-sided ataxia while reading the newspaper on the morning on admission. This is on a background of hypertension and dyslipidemia. Examination on presentation indicated a left internuclear ophthalmoplegia with a left sixth cranial nerve palsy. Past pointing was noted on the left arm, and corresponding abnormal heel–shin test was identified on the left leg. Mild right arm weakness was also noted, and sensory examination was normal. Notably heart sounds were dual with no murmurs.

He was thrombolysed within 2 h of symptom onset. His computed tomography (CT) angiogram did not show any basilar or vertebral occlusion or dissection. Magnetic resonance imaging (MRI) of the brain indicated diffusion restriction in the left medial midbrain and left cerebellar regions consistent with clinical findings (Figs. 1 to 5). There was no evidence of atrial fibrillation on ECG or telemetry.

Transthoracic echocardiogram indicated a 24×18×20-mm isoechoic, solid, highly mobile mass attached to the anterior left atrial wall below the anterior mitral leaflet. Transesophageal echocardiogram confirmed the presence of a spherical mobile mass attached to the orifice of the left atrial appendage (Figs. 6 and 7, Supplementary Video 1 “Supplementary video 1: TOE video.avi”).

Our patient was transferred to a tertiary center for resection. Under cardiopulmonary bypass and cardioplegic arrest, a fragile 22-mm round pedunculated mass was excised from the ostium of the left atrial appendage. The left atrial appendage was then oversewn. An incidental patent foramen ovale (PFO) was noted intraoperatively and also oversewn. Coronary artery bypass was performed for coincidental coronary artery disease.

Histopathologic analysis of the 22×18×12-mm ovoid mass however noted branching of papillary fronds lined with epithelial cells and collagenous core with focal areas of myxoid change consistent with a papillary fibroelastoma (Figs. 8 and 9).

Our patient's complex ophthalmoplegia resolved by day 3 into admission; however, his left cerebellar ataxia persisted and required rehabilitation. After 3 months postexcision, there have been no further cerebrovascular events.

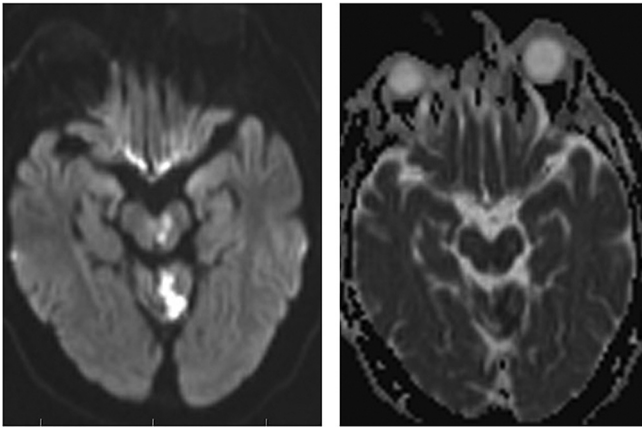
## 2. Discussion

CPFs are benign primary cardiac tumors and a rare but increasingly recognized cause of cardioembolic stroke and transient ischemic attack. While previously thought to be less common than atrial myxomas, with increasing echocardiology surveillance, CPFs are now the most common benign cardiac tumor [1,2]. CPFs commonly have a predilection for aortic and mitral valves [3] and are rarely tethered to the atrial epithelium. Left atrial CPFs are exceedingly rare, estimated to be between 1.3% and 6.1% of all CPFs as described by large retrospective studies [3–6]. Unlike atrial myxomas, they have not been reported to present with a systemic prodrome [1]. A recent retrospective analysis of 67 cases of stroke in CPF demonstrated anterior circulation infarcts in 93% of cases [7].

The authors report no conflict of interest.

\* Corresponding author.

E-mail address: ljbbonavia@gmail.com (L. Bonavia).

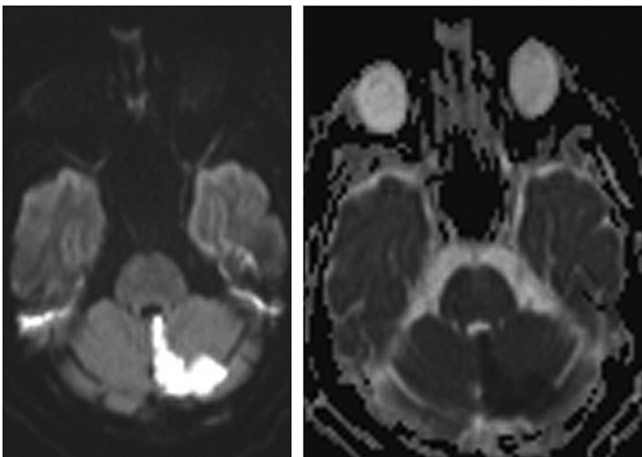


**Figs. 1 and 2.** MRI diffusion-weighted imaging and corresponding ADC image of ischemia within midbrain and base of cerebellum.

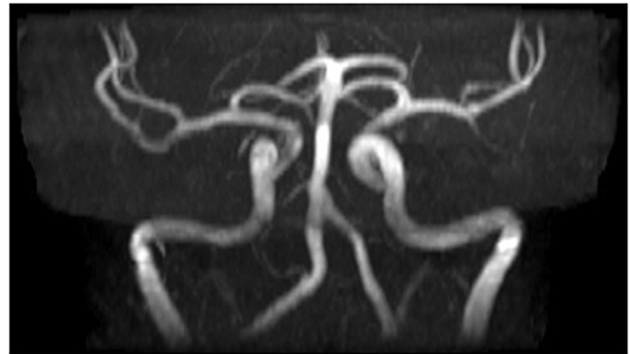
They have fragile, frond-like morphology and a collagenous core and are pedunculated in about half of cases [3,8]. When submerged in saline, they resemble sea anemones [9]. Of the few cases of left atrial CPFs described, they were resected from the superior and inferior aspects of the pulmonary veins, atrial appendage, and septum, and one case tethered to the right atrium [9–14]. They average between 10 mm and 30 mm in diameter and can be identified on echocardiography [5]. Sensitivity and specificity of transthoracic echocardiography, however, reduce with sizes less than 20mm<sup>3</sup>. Echocardiographic workup should therefore include transesophageal echocardiography in suspected cases.

Unlike atrial myxomas, CPFs have a mean age of 60 years at diagnosis without a predominant gender [1,3]. Out of the cases of left atrial CPF, patient ages ranged from 55 to 80 [9,12–17]. However, valvular CPFs have been reported to occur in patients less than 30 years of age [18,19]. Posterior circulation infarct from a left atrial CPF has been described once [14]. A recent retrospective analysis of 67 cases of stroke in CPF demonstrated anterior circulation infarcts in 93% of cases [7].

CPFs are thought to be acquired rather than inherited, with possible contributing factors including epithelial hyperproliferation from hemodynamic trauma, organized thrombi, and underlying genetic predisposition [3]. Prior cardiac procedures, radiation therapy, and chronic CMV endocarditis have also been proposed as contributing factors [3]. None of these factors are present in our case. Given their fragile composition, segments of the tumor may dislodge and contribute to systemic embolization. Additionally, the mucopolysaccharide and hyaluronic-acid-rich frond surface may harbor thrombi that may embolize. In a retrospective study



**Figs. 3 and 4.** MRI diffusion-weighted imaging and corresponding ADC image of ischemia within cerebellum.

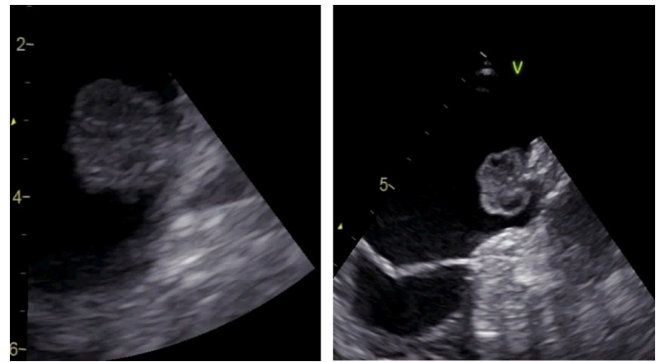


**Fig. 5.** Normal MRI angiogram vessel construct.

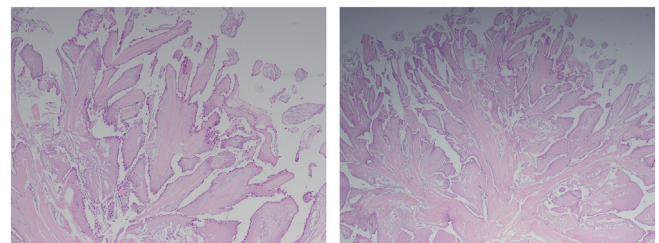
of 725 cases of CPF, thrombi were noted on 19 cases perioperatively, and 3 cases of recurrent strokes were noted despite warfarinization [20].

Large retrospective analyses estimate stroke or transient ischemic attack risk in CPF to be between 13.5% and 53.6% [2,4–6,8,20]. Recurrence of stroke in nonresected tumors at diagnosis was estimated to be between 16% and 24% with mean follow-up of 31 months [5,20]. Sun et al., however, noted a recurrence rate of over 50% with a median follow-up of 130 days [8]. Tamin et al. recently demonstrated a recurrence of stroke in nonresected CPF to be 6% in 12 months and 13% in 5 years [2]. Cases of stroke due to valvular or nonvalvular CPF have been reported to respond to thrombolysis, such as this case [7]. After resection, stroke recurrence rates are low [2,4,8], and survival rates are excellent [21]. However, it is important to note that while stroke recurrence is significantly reduced after resection, background cerebrovascular risk warrants consideration [2]. There is no consensus on appropriate anticoagulation postresection, although anticoagulation is warranted if surgery is not an option [3].

It is important to identify that our patient does have known cerebrovascular risk factors of hypertension and dyslipidemia [22]. However,



**Figs. 6 and 7.** Atrial views of the mobile appendicular mass noted on transesophageal echocardiography.



**Figs. 8 and 9.** Histopathology of the ovoid mass at 2× and 4× magnification. Note the frond-like appearance, peripheral epithelial cells, and collagenous core (thanks to Dr. Dhaval Joshi MBBS FRCPA, Melbourne Pathology, Australia).

the multiembolic nature of the infarcts suggests a cardiac or proximal arterial source. A normal CT and MRI angiogram makes a proximal arterial source unlikely (Fig. 5).

Furthermore, a concomitant PFO was identified intraoperatively. PFOs are found in 40% of the population, and risk of stroke is not well established [23]. Given the presence of a CPF with its recognized embolic stroke risk as described above, it is more likely that our patient's stroke was attributable to the identified atrial source rather than his cerebrovascular risk factors alone or his PFO.

This case highlights the growing recognition of CPF as a rare but important cause of cardioembolic stroke in the older population. Differential diagnosis of a left atrial mass should include a CPF. The risk of stroke recurrence for a given diagnosis is high, and prompt surgical resection is recommended.

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.carpath.2019.01.007>.

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