Multiple causes for an ischemic stroke: myxoma, papillary fibroelastomas and patent foramen ovale

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HSR Proceedings in Intensive Care and Cardiovascular Anesthesia 2012; 4(3): 187-191

ABSTRACT

We report a case in which multiple uncommon causes of an ischemic vascular accident coexisted in the same patient. The patient was admitted with signs of acute stroke. Investigation workup revealed a left atrial tumor (myxoma) and a patent foramen ovale. Intraoperatively, transesophageal echocardiography added new information: papillary fibroelastomas were found in the aortic valve. This finding dictated a change in the surgical plan, adding resection of aortic valve masses to the planned excision of the left atrial tumor and patent foramen ovale closure. The uniqueness of this case derives from the coexistence of rare primary cardiac tumors. There are only five cases in literature of myxoma concomitant with fibroelastoma and the occurrence of multiple fibroelastoma is also extremely rare. Moreover this case emphasizes the benefit of the intraoperative use of transesophageal echocardiography to improve the diagnosis and management of cardiac surgical patients.

Keywords: *papillary fibroelastoma, myxoma, patent foramen ovale, stroke, intraoperative transesophageal echocardiography, embolism.*

CASE REPORT

A 64 year-old woman was admitted with right arm weakness. Brain computed tomography was consistent with ischemic cerebral vascular accident, showing two small ischemic lesions on the left hemisphere, one in the head of the caudate nucleus and the other in the white matter of the semi-ovale center.

There was no record of previous infarcts in the cerebral territory or in other organs or peripheral regions.

Besides arterial hypertension, controlled with an agiontensin converting enzyme inhibitor, there were no other known risk factors such as smoking, diabetes or dyslipidemia.

Other than the patient's father who had died of stroke at a late age, no other family risk factors could be identified. Carotid/vertebral echodoppler was normal. Transthoracic echocardiography and transesophageal echocardiocardiography (TEE) revealed a left atrial mass.

The mass was described as spherical, heterogeneous, with a size of 29x23x32 mm,

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mobile, attached by a short stalk to the posterior wall. Mitral and aortic valves were normal and left ventricular function was preserved. TEE also reported the presence of a patent foramen ovale (PFO).
The patient was scheduled for surgery. Intraoperative TEE confirmed the presence of the previously reported lesions (left atrial mass and PFO) but also revealed an abnormal aortic valve.
A nodular, oval mass (9x7 mm) was present in the non-coronary leaflet and valvular excrescences (around 10 mm length) arose from both the non- coronary and the right coronary leaflets and projected into the aorta. These multiple frond-like masses were highly mobile.
Surgery was performed under hypothermic (32°C) cardiopulmonary bypass. After aortic cross-clamping, anterograde cardioplegia was performed. Through left atriotomy a grey gelatinous tumor was found attached to the superior aspect of the posterior wall of the left atrium, near the ostium of the left superior pulmonary vein. This left atrial mass was resected.

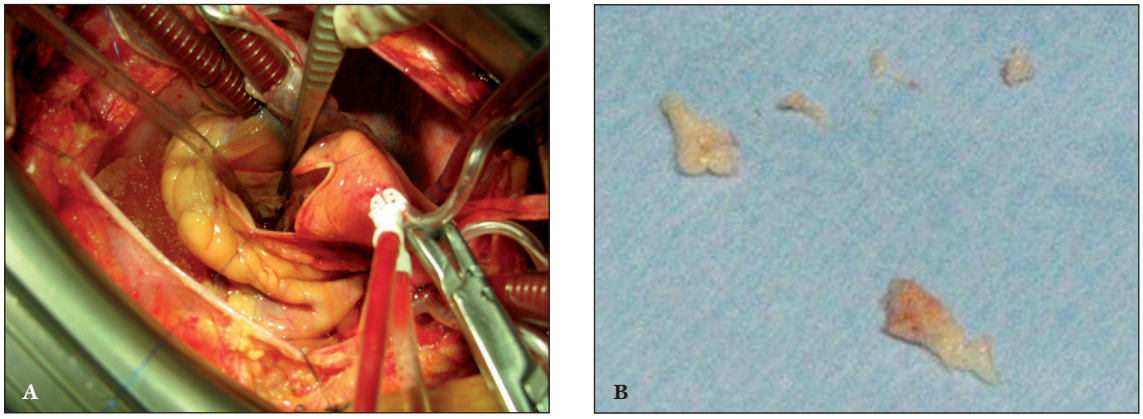

Figure 1 - *Aortic valve papillary fibroelastomas in situ (A) and after resection (B).*

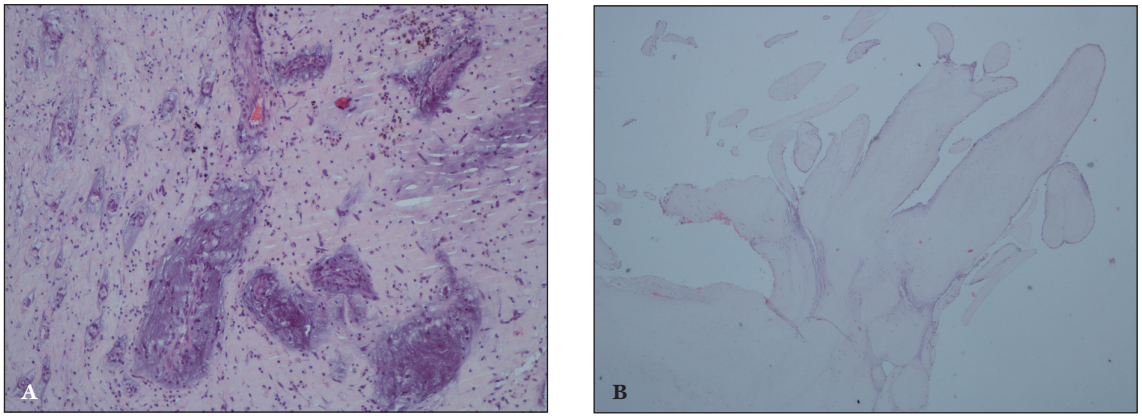

Figure 2 - *A - Myxoma (Hematoxylin-Eosin x100): neoformation consisting of mucous pools and eosinophilic loose connective tissue stroma with low celularity, mainly fibroblasts and inflammatory cells.*
B - Aortic valve papillary fibroelastoma (Hematoxylin-Eosin x40): exophytic neoformation consisting of several digitiform processes with loose connective tissue axis covered by a thin endotelial layer.

In order to assure better visualization and by surgical option, PFO was sutured via right atriotomy. Inspection of the aortic valve revealed the presence of two nodular formations, with multiple villous projections, attached to either the right or the non-coronary cusps. These abnormal structures were resected while sparing the valve structure (*Figure 1*).
Weaning from bypass was easy without the need of inotropic support. Postoperative course was uneventful. No adverse events or complications took place during the 3 months follow up.
Pathological findings of the resected masses were consistent with left atrial myxoma and aortic valve papillary fibroelastomas (PFE) (*Figure 2*).

DISCUSSION

Cerebrovascular ischemia is generally the result of atheroemboli from a central vessel or in situ thrombosis of a cerebral vessel or emboli from a cardiac source (usually left chamber thrombi). A minority of ischemic strokes is associated with other sources of cardioembolism: PFO is the most prevalent among these, while primary cardiac tumors are rare.
PFO is present in 27 % of adults. Its persistence allows for venous thromboemboli to enter the arterial circulation. It has been identified as a potential risk factor for stroke, by means of paradoxical embolism, and it is present in more than 50 % of young patients with cryptogenic stroke (1).
Primary cardiac tumors of the heart are rare. The estimated incidence is approximately 0.02 %, in autopsy series. Myxoma is the most frequent primary cardiac tumor and its clinical presentation may vary from an asymptomatic finding to sudden death. Common presentations include fatigue, chest pain, pulmonary edema and infarcts: cerebral, myocardial and peripheral. Cardiac myxoma is predominantly a left atrial tumor that is usually attached to the atrial septum by a stalk. PFE is the third most common primary cardiac tumor (after myxoma and lipoma) accounting for approximately 7 % of cases. It is the most common cardiac valve tumor. It may be found anywhere in the heart, however approximately 80 % are found on the valvular endocardium (predominantly on the aortic valve).
Usually it presents as a small, solitary, pedunculated and mobile tumor (2). Manifestations include transient ischemic attacks, stroke, myocardial infarct, peripheral infarct, syncope and even sudden death. Both these tumors are benign in nature, but, regardless of their size, they are potential embolic sources to the brain and the coronary circulation.
The sources of cardioembolic stroke usually occur isolated. However, in the present case, multiple causes coexisted in the same patient, any of which might have been the cause of the described cerebral ischemic event. Some interesting points of this case report need to be emphasized:

1) Combined presence of histologically different primary cardiac tumors.

The probability of a patient developing either a cardiac myxoma or a PFE during a lifetime is very low.
The probability of developing both tumors simultaneously is even lower. Concomitant tumors have been infrequently reported, with only five cases documented (3-7). Prifti et al. (3) describe a case of myxoma on the atrial surface of the mitral anterior leaflet coexisting with a papillary fibroelastoma on the leaflet´s ventricular aspect.
Menon et al. (4) reported the case of a myxoma in the fossa ovalis and a PFE on the posterior wall of the left atrium. Jutley et

al. (5) described the presence of a myxoma on the lateral wall of the right atrium and a PFE on the left atrium surface of the inter-atrial septum. Matsushita et al. (6) also reported a case of concomitant left atrial myxoma and aortic valve PFE. Agaimy et al. (7) described the case of a myxoma (implanted on the left atrium aspect of the inter-atrial septum) with several areas of PFE-like tissue.

2) Existence of multiple PFE.

PFE are usually single tumors, multiple tumors are extremely rare (only 5 % of PFE present as multiple tumors).

Usually they appear at different locations in the heart (multiple intraventricular sites or involving separate cardiac valves). Very few cases of multiple tumors on the aortic valve have been published in the literature (8-11).

3) Incidental detection of PFE during intraoperative TEE.

Before echocardiography was available, PFE were usually only found by chance during cardiac surgery or autopsy.

The advent of echocardiography and the routine use of intraoperative TEE in cardiac surgery are responsible for an increasing number of reported cases of PFE.

4) Intraoperative TEE examination changed the surgical plan.

This case report shows quite clearly the importance of a complete intraoperative TEE examination.

Intraoperative TEE can be more detailed and comprehensive than pre-operative TEE because it is performed under general anesthesia.

Often the pre-operative TEE is a focused examination as a complete examination is not always possible (for example in the case of an awake, non cooperative patient).

It is a well-known fact that intraoperative TEE can reveal new information, which is why its routine use is strongly recommended by many authors. In our center we perform intraoperative TEE on a routine basis.

Though PFE are asymptomatic in the majority of patients, their identification is important because (due to their embolic risk) these tumors may result in life-threatening complications.

The fact that in our case report the aortic valve presented a multiple PFE obviously increased the risk of embolic events. Identification of these tumors allowed for surgical excision that was curative since recurrence of PFE has not been reported.

In summary, our case highlights a rare combination of PFO and histologically different primary cardiac tumors (left atrial myxoma and two aortic PFE), any of which could have been the cause of the cerebral ischemic event that the patient was admitted for.

The probability of a patient developing a myxoma and multiple PFE simultaneously is extremely low. The coexistence of a PFO (even though it is relatively common) adds to the rarity of our case.

The PFE were incidentally found by intraoperative echocardiography, a fact that reinforces the importance of its use.

TEE is now used routinely during cardiac surgery in many centers; intraoperative TEE allowed for the correction of all surgically treatable causes of cardiac embolism present in this patient during the same surgery.

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Cite this article as: Roque J, Silva F, Arruda Pereira R, Cravino J. Multiple causes for an ischemic stroke: myxoma, papillary fibroelastomas and patent foramen ovale . HSR Proceedings in Intensive Care and Cardiovascular Anesthesia 2012; 4(3): 187-191.

Source of Support: Nil. **Conflict of interest:** None declared.