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CASE REPORT

Aortic Valve *Myxoma* Presenting with a Stroke

A case report and review of the literature

*Feras A. Alkuwaiti,[1] Yasser Elghoneimy,[2] Sami Ghazal[3]

ورم مخاطي في الصمام الأبهري يعرض كسكتة دماغية

تقرير حالة و مراجعة للأدبيات الطبية

فراس أحمد الكويتي، ياسر الغنيمي، سامي غزال

ABSTRACT: *Myxomas* originating from the aortic valve are rare. We report a 40-year-old male patient who presented to the King Fahd Hospital of the University, Khobar, Saudi Arabia, in 2017 with a stroke. Transoesophageal echocardiography indicated a mobile mass measuring 6 × 2 mm attached to the right coronary cusp of the aortic valve and a mobile interatrial *septum* with a small patent *foramen ovale* (PFO). The patient underwent surgical excision of the mass and direct closure of the PFO. Histopathology confirmed the mass to be a *myxoma*. Despite their rarity, the recognition and treatment of valvular *myxomas* is very important; moreover, clinicians should be aware that affected patients may present with an embolic stroke.

Keywords: Aortic Valve; Myxoma; Patent Foramen Ovale; Stroke; Transesophageal Echocardiography; Case Report; Saudi Arabia.

الملخص: يعد الورم المخاطي الناشئ عن صمامات القلب حالة نادرة الحدوث. نعرض هنا حالة مريض رجل يبلغ من العمر 40 عامًا تقدم إلى مستشفى الملك فهد الجامعي، الخبر، المملكة العربية السعودية، في عام 2017 بسكتة دماغية. أشار تخطيط صدى القلب عبر المريء إلى وجود كتلة متحركة بحجم 2 × 6 مم متصلة بالشرفة التاجية اليمنى للصمام الأبهري والحاجز الأذيني مع وجود ثقب بيضاوي قلبي صغير. خضع المريض للإستئصال الجراحي وإغلاق مباشر للثقب البيضاوي القلبي. أكد تقرير مرضيات الأنسجة على أن الكتلة هي عبارة عن ورم مخاطي. على الرغم من ندرة الحالة، إلا أن معرفة وعلاج الورم الحبيبي للصمامات يعتبر مهمة للغاية؛ علاوة على ذلك، يجب أن يكون الأطباء على دراية بأن المرضى المصابين قد يظهرون بسكتة دماغية صميّة.

الكلمات المفتاحية: الصمام الأبهري؛ ورم مخاطي؛ الثقب البيضاوي القلبي؛ السكتة الدماغية؛ تخطيط صدى القلب عبر المريء؛ تقرير حالة؛ المملكة العربية السعودية.

Myxomas originating from the cardiac valves are uncommon.[1,2] These benign tumours typically occur between the fourth and seventh decades of life and usually affect women more commonly than men.[1] The condition is mostly sporadic in nature, but can also be familial.[2] *Myxomas* are true neoplasms in that they are derived from primitive multipotent mesenchymal cells which may exist as embryonic remnants in the heart wall.[3]

Rarely, *myxomas* may arise from both the ventricular aspect and the margin of the aortic valve cusps; moreover, one or both leaflets may also be affected.[4] Although transoesophageal echocardiography (TEE) aids in the assessment of such masses, histopathology is considered the gold-standard method to confirm a diagnosis.[4,5] This report describes an atypical case of aortic valve *myxoma* and patent *foramen ovale* (PFO) presenting with a cerebral stroke. Such cases serve to increase awareness of the varying presentation of aortic valve *myxomas* and highlight appropriate investigations and management of this condition.

Case Report

A 40-year-old male patient presented to the King Fahd Hospital of the University (KFHU), Khobar, Saudi Arabia, in 2017 with sudden-onset dysphasia, left *ataxia*, nausea and vomiting over the preceding two hours. The patient reported having had a previous stroke at the age of 11 years; however, he was unable to provide any information about the event. At presentation, the patient was alert, oriented and had stable vital signs. All laboratory investigation findings were within normal limits and electrocardiography showed normal sinus rhythm. As a result, the case was transferred to the stroke team. Urgent computed tomography (CT) of the head showed evidence of the prior ischaemic stroke event in the right occipital lobe and right cerebellum. An intravenous thrombolytic was administered in accordance with hospital guidelines. Subsequently, the patient was admitted to the inpatient department and received 150 mg of aspirin and 75 mg of clopidogrel. Symptoms improved over the course of the following three days. However,

Departments of [2]Surgery and [3]Internal Medicine, [1]College of Medicine, Imam Abdulrahman Bin Faisal University, Dammam, Saudi Arabia
**Corresponding Author's e-mail: feras_a_k@yahoo.com*

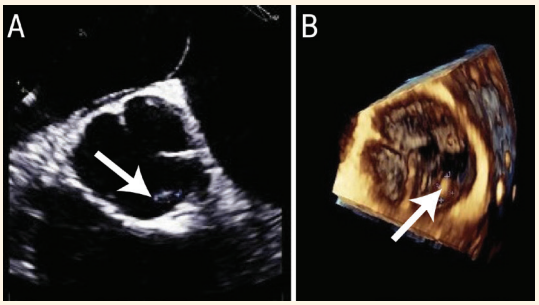

Figure 1: Transoesophageal echocardiography of a 40-year-old male stroke patient in (**A**) bi-plane short-axis view showing a small mass in the right coronary cusp of the aortic valve measuring 3 × 5 mm and (**B**) three-dimensional enfaced view showing the mass measuring 6 × 2 mm.

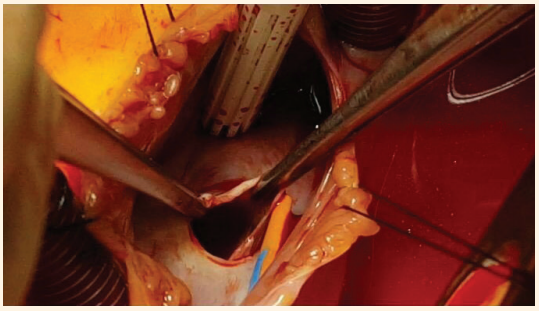

Figure 2: Intraoperative photograph of the heart of a 40-year-old male stroke patient showing an opening between the right and left *atria* measuring 10 × 10 mm.

further investigations were undertaken to determine the source of the stroke.

Brain magnetic resonance imaging (MRI) revealed that the patient had suffered an acute ischaemic stroke, with a multifocal area of diffusion restriction within the right cerebral and superior cerebellar peduncles and both cerebellar hemispheres, as well as hyperintensities on T2-weighted fluid-attenuated inversion recovery MRI sequences. However, transthoracic echocardiography (TTE) did not show any clear source of *emboli*. Accordingly, TEE was performed and revealed a small mobile mass measuring 6 × 2 mm attached to the right coronary cusp (RCC) of the aortic valve and a mobile interatrial *septum* with a small PFO visible with agitated saline contrast [Figure 1]. No significant arrhythmia was noted on a Holter monitor. A papillary fibroelastoma (PFE) was suspected.

After two weeks of stroke management, the decision was made excise the mass along with closure of the PFO to prevent any future embolic events. The surgical procedure was performed while the patient underwent cardiopulmonary bypass. During the surgery, the PFO was visible as an opening between the right and left *atria* [Figure 2]. The mass was successfully excised from the RCC and the PFO was closed. The postoperative period was uneventful and the patient was discharged five days later. A histopathological examination of the excised mass showed a paucicellular lesion composed of stellate and oval cells with a moderate amount of eosinophilic cytoplasm, fine chromatin and indistinct cell borders [Figure 3]. These features were consistent with a diagnosis of *myxoma*.

Discussion

To the best of the authors' knowledge, only 11 cases of aortic valve *myxoma* have been previously reported in the literature [Table 1].[4–14] Interestingly, nine of these previously reported patients were male, as in the current case; this is contrary to the usual female preponderance of *myxoma* cases.[1,4–9,11,13,14] Overall, six aortic valve *myxoma* patients were symptomatic, with symptoms largely attributable to distal embolisation and *ischaemia*, including three cases with stroke, two with myocardial infarctions and one with limb *ischaemia*.[4,6,8,11–13] Clinicians should therefore be aware of stroke as a potential presentation of aortic valve *myxoma*.

The pathogenesis of ischaemic events in aortic valve *myxoma* could be due to either embolic debris derived from the tumour itself or from a *thrombus* around the tumour.[12] In the current case, the patient was also diagnosed with PFO, a potential cause of embolic stroke in which a paradoxical embolism may pass through a PFO via right-to-left shunting (RLS).[15] However, in the current case, the observed PFO was very small in size and therefore would not be able to cause the RLS of an *embolus*.

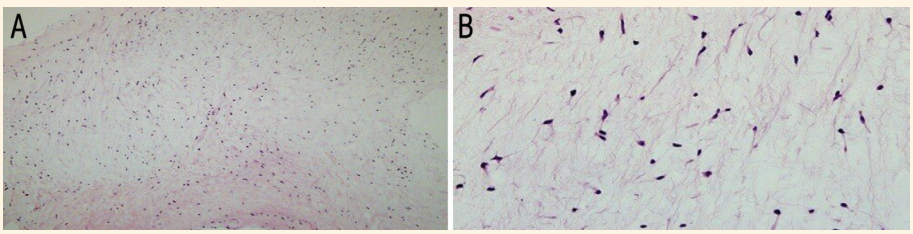

Figure 3: Haematoxylin and eosin stains at (**A**) x40 magnification showing a hypocellular lesion in a myxoid matrix and (**B**) at x200 magnification showing stellate and oval cells with eosinophilic cytoplasm and indistinct cell borders.

Table 1: Literature review of reported aortic valve *myxoma** cases[4–14]

Author and year of case*	Age/ gender	Presentation	History	Imaging	Site	Size in cm	Complication	Surgery
Kennedy *et al.*[8] (1995)	23/M	Leg pain	None	TTE	RCC and LCC	1.5	Limb *ischaemia*	AVR
Watarida *et al.*[9] (1997)	58/M	Asymptomatic	None	TTE	RCC	1.1 × 1	None	AVR
Ramsheyi *et al.*[6] (1998)	34/M	Facial haemiparesis	None	TTE and TEE	RCC	1	Stroke	AVR
Okamoto *et al.*[10] (2006)	61/F	Endocarditis	HTN and DM	TTE	LCC	1 × 1	None	Resection
Dyk *et al.*[11] (2009)	15/M	Chest pain	None	TTE and TEE	NCC	4 × 1	MI	Resection
Koyalakonda *et al.*[12] (2011)	60/F	Paroxysmal AF	AF and HTN	TTE and TEE	RCC	1 × 1	Stroke	Resection
Fernández *et al.*[13] (2012)	28/M	Haemiparesis	Epilepsy	TTE	RCC and LCC	1.5 × 0.7	Stroke	AVR
Kim *et al.*[7] (2012)	72/M	Shortness of breath	HTN	TTE	NCC	1.5 × 0.8	None	AVR
Javed *et al.*[4] (2014)	81/M	Leg pain	HTN, CKD and PAD	CT, angiography and TEE	LCC	1.8 × 1.2	Acute MI	CABG and resection
Prifti *et al.*[5] (2015)	13/M	Dyspnoea and *angina*	None	TTE and TEE	RCC and LCC	0.6 × 0.2	None	AVR
Ji *et al.*[14] (2017)	17/M	Heart murmur	None	TTE	NCC	2	None	AVR
Present case (2018)	40/M	Dysphasia, *ataxia* and nausea	Previous stroke	TTE and TEE	RCC	0.6 × 0.2	Stroke and PFO	Excision and PFO closure

*TTE = transthoracic echocardiography; RCC = right coronary cusp; LCC = left coronary cusp; AVR = aortic valve replacement; TEE = transoesophageal echocardiography; HTN = hypertension; DM = diabetes mellitus; NCC = non-coronary cusp; MI = myocardial infarction; AF = atrial fibrillation; CKD = chronic kidney disease; PAD = peripheral arterial disease; CT = computed tomography; CABG = coronary artery bypass graft; PFO = patent foramen ovale. *In all cases, the diagnosis was confirmed via histopathological examination.*

The differential diagnosis of an aortic valve *myxoma* includes vegetation, a *thrombus*, PFE and Lambl's excrescences; these conditions can be distinguished by their microscopic and immunohistochemical characteristics.[7,10,13] In addition, *thrombi* and *myxomas* can be differentiated using two-dimensional echocardiography; the former typically present with a layered appearance, while an area of echolucency may be observed within the tumour in the latter.[7] Moreover, vegetations of the heart valves are a sign of infective endocarditis; however, this was ruled out in the current case due to a lack of history of this condition.[10,16]

On imaging, the size, shape, attachment site, mobility and location of a cardiac mass can aid in the exact diagnosis, along with the patient's clinical presentation.[17–19] For example, in PFE cases, the tumour appears as a small mass attached to the mitral or aortic valve, usually located downstream and attached by a small pedicle; such masses are more common and often mobile and appear irregularly-shaped with delicate frond-like surfaces.[7,17,18] In contrast, Lambl's excrescences appear as filiform fronds located near valve closure lines.[20] Radiologically, TEE is superior to TTE in identifying a cardiac mass and defining its exact location, attachment to the underlying tissue and relation to surrounding structures.[21,22] Among previously reported cases of aortic valve *myxoma*, 10 were identified by TTE, of which four were further confirmed via TEE.[5–14] In one case, an aortic valve abnormality was first revealed via CT.[4] In the current case, TEE revealed the mass to be located on the RCC of the aortic valve, thus precluding a *thrombus* or cardiac vegetation; however, the preoperative diagnosis was of a PFE. As with previous cases, the correct diagnosis was eventually confirmed by histopathological examination revealing a myxoid matrix with ovoid *myxoma* cells.[4–14]

Typically, the management of aortic valve *myxoma* cases involves the surgical excision of the tumour as well as the surrounding tissue to minimise the risk of local recurrence, as in the present case.[4,10–12] During the procedure, the native aortic valve should be conserved as much as possible; however, if the tumour is too large

and/or degeneration of the valvular structure occurs, aortic valve replacement (AVR) may be necessary. Among previously reported cases, seven underwent AVR, while resection alone was sufficient in three cases and one required resection and a coronary artery bypass graft.[4–14] Nevertheless, regardless of treatment option, aortic valve *myxoma* cases require careful follow-up due to the high probability of local recurrence and distal tumour growth at the site of embolisation.[5]

Conclusion

An aortic valve *myxoma* is uncommon and may present with serious complications, such as stroke. In patients with embolic phenomena, TEE should be performed, with special attention to the valvular morphology. For valvular *myxomas*, aggressive surgical excision is necessary to prevent structural valve degeneration and local recurrence.

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