The role of intraoperative transoesophageal echocardiography in the diagnosis and management of a rare multiple fibroelastoma of aortic valve: a case report and review of literature

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Papillary fibroelastoma is the third most common primary tumour of the heart that usually involves the cardiac valves. Multiple papillary fibroelastomas are extremely rare. We report a case with multiple valve papillary fibroelastoma which was identified only by intraoperative transoesophageal echocardiography. The patient complained of atypical chest pains. She was affected by coronary artery disease and had previously had a myocardial infarct. This finding dictated a change in the operative approach. The aortic valve resection was performed in addition to coronary revascularization. If the intraoperative transoesophageal echocardiography was not performed, our patient would have had just coronary artery bypass graft surgery, probably without solving the symptoms. Furthermore, in future she would have undergone another cardiac operation for resection of aortic masses and valve replacement. The intraoperative use of Transoesophageal Echocardiography improves the diagnosis and the management of all cardiac surgical patients.

KEYWORDS
Multiple fibroelastoma; Aortic valve; Intraoperative transoesophageal echocardiography; Coronary artery bypass graft surgery

Case report
A 59-year-old woman was hospitalized in our Department to be submitted to coronary artery bypass graft surgery (CABG).

She had dyslipidaemia and systemic arterial hypertension as risk factors for coronary artery disease.

Her anamnestic history revealed, 4 months before, a myocardial infarct—STEMI—of the inferior wall. It was treated with primary Percutaneous Transluminal Coronary Angioplasty and two drug-eluting stents were implanted on the right coronary artery. One month later, she underwent a new coronary angiography for a new and transient episode of chest pain. Two other drug-eluting stents were implanted on the left anterior descending artery and on the right coronary for the evolution of the atherosclerotic disease. However, intravascular ultrasound documented a stenosis of the left coronary bifurcation (approximately 55%) but cardiac surgery was not considered useful.

After another month, the patient experienced numerous and brief atypical chest pains; they spontaneously resolved after few minutes. Therefore, the patient was run to CABG.

During admission in our Department, the physical examination did not show abnormalities. Myocardial necrosis markers and D-dimer were negative. Blood pressure was 140/80 mmHg. Heart rate was 64 bpm in sinus. Atrium-ventricular and intra-ventricular conduction were normal. Electrocardiographic changes in inferior peripheral leads suggested a previous myocardial infarct. Echocardiography revealed that the left ventricular chamber had normal volume and wall thickness. Ejection fraction was preserved (55%) despite hypokinesia of inferior wall and a remodelling in the same area. The right chambers were normal. Mitral valve was fibrotic with a mild regurgitation; the aortic valve, instead, showed a thickening and fibrosis of leaflet with moderate incontinence. A new angiography confirmed

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the stenosis of approximately 55% at the left coronary bifurcation. Intra-surgery transoesophageal echocardiography was performed and an incidental lesion of the aortic valve was found (Figure 1; see Supplementary data online, Videos S1, S2, S5). The two coronary cusps showed multiple frond-like masses (Supplementary data online, Video S3) with high-frequency oscillations during cardiac cycle. This finding dictated a change in the operative approach and surgeon tried to resect the two valvular masses (Figure 2). In spite of the resection, aortic regurgitation increased. For this reason, the entire aortic valve needed to be replaced by a mechanical prosthesis (St. Jude 21). Coronary revascularization was performed along with CABG. Large safena vein was grafted to the first obtuse marginal branch and the left internal mammary artery to the left anterior descending artery. After prosthesis implant, the examination of the native aortic valve showed two masses of approximately 5 × 5 mm. The first was on the ventricular side of left cusp and the other one attached to the edge of the right cusp. The Cardiovascular Pathology Unit received the aortic leaflets and the papillary neoplasia. At histology, the leaflets showed a normal architecture with expansion of the spongiosa, where a collection of glycosaminoglycans was apparent. The fibrosa was thinned and the free margin of the valve exhibited a rolled edge, as a consequence of the mechanical effect of regurgitation. The papillary tumour was made of slender papillae with an elastic core, corresponding to the classic histological features of a papillary fibroelastoma (Figure 3). No thrombotic material was apparent in between the neoplastic papillae and this observation supports the hypothesis that the coronary embolism derives from the detachment of some villous projections and that it is not thrombotic in origin.

After cardiac surgery, the patient had no complaints of atypical chest pain.
Cardiac papillary fibroelastoma is a rare cardiac neoplasm predominantly affecting cardiac valves. Edwards and colleagues at the Armed Forces Institute of Pathology reported that papillary fibroelastomas made up nearly 75% of the cardiac valve tumours found during their multi-institutional study that spanned six decades. Most papillary fibroelastomas reported in literature have been found by chance on surgical excision of mitral or aortic valves. Others have been identified during autopsies. Recently, the use of trans-thoracic and transoesophageal echocardiography has allowed to identify these tumours also in vivo. Half of these are symptomatic for coronary embolism and may present worse complications: cerebral embolism, acute valvular dysfunction, and sudden death. At pathological examination these tumours are small, benign, solitary neoplasms, and rarely multiple. Although they commonly affect cardiac valves, they sometimes occur on the chordae tendineae, on papillary muscles, on interventricular septum, on ventricular outflow tract. Nowadays, the origin of these tumours is still considered unknown. Most authors identify them as hamartomas; other ones, as Marvasti et al., believe they are true neoplastic growths associated with chronic trauma. In the present case report, the authors postulate, as Eckstein et al. have reported, that the previous inferior wall infarction may have been caused by papillary fibroelastoma of the aortic valve. Embolization may have occurred from papillary fronds or alternatively, as suggested by Topol et al., these tumours may have provided a nidus for platelet and fibrin aggregation, widely accepted as a cause of thrombotic embolization. This last hypothesis seems not to apply to our case since the patient received a daily double antiaggregant therapy following the implanted medicated stents. It is then possible to presume that embolization of little pieces of papillary fronds caused the persistent symptomatology we have just described. It is important to remind that in this case the valve presented a multiple papillary fibroelastoma, which De Virgilio et al. or Eslami and coworkers described as a rare affection of the heart. The fact that it was multiple obviously increased the risk of embolic events. Our case is also interesting owing to the fact that the valve also exhibited a collection of glycosaminoglycans: it is then possible to presume that, while the papillary fibroelastoma have been causing the frequent episodes of embolization, this collection of glycosaminoglycans is to be considered the majority responsible for the valve incompetence (Supplementary data online, Videos S4, S5).

**Discussion**

Cardiac papillary fibroelastoma is a rare cardiac neoplasm predominantly affecting cardiac valves. Edwards and colleagues at the Armed Forces Institute of Pathology reported that papillary fibroelastomas made up nearly 75% of the cardiac valve tumours found during their multi-institutional study that spanned six decades. Most papillary fibroelastomas reported in literature have been found by chance on surgical excision of mitral or aortic valves. Others have been identified during autopsies. Recently, the use of trans-thoracic and transoesophageal echocardiography has allowed to identify these tumours also in vivo. Half of these are symptomatic for coronary embolism and may present worse complications: cerebral embolism, acute valvular dysfunction, and sudden death. At pathological examination these tumours are small, benign, solitary neoplasms, and rarely multiple. Although they commonly affect cardiac valves, they sometimes occur on the chordae tendineae, on papillary muscles, on interventricular septum, on ventricular outflow tract. Nowadays, the origin of these tumours is still considered unknown. Most authors identify them as hamartomas; other ones, as Marvasti et al., believe they are true neoplastic growths associated with chronic trauma. In the present case report, the authors postulate, as Eckstein et al. have reported, that the previous inferior wall infarction may have been caused by papillary fibroelastoma of the aortic valve. Embolization may have occurred from papillary fronds or alternatively, as suggested by Topol et al., these tumours may have provided a nidus for platelet and fibrin aggregation, widely accepted as a cause of thrombotic embolization. This last hypothesis seems not to apply to our case since the patient received a daily double antiaggregant therapy following the implanted medicated stents. It is then possible to presume that embolization of little pieces of papillary fronds caused the persistent symptomatology we have just described. It is important to remind that in this case the valve presented a multiple papillary fibroelastoma, which De Virgilio et al. or Eslami and coworkers described as a rare affection of the heart. The fact that it was multiple obviously increased the risk of embolic events. Our case is also interesting owing to the fact that the valve also exhibited a collection of glycosaminoglycans: it is then possible to presume that, while the papillary fibroelastoma have been causing the frequent episodes of embolization, this collection of glycosaminoglycans is to be considered the majority responsible for the valve incompetence (Supplementary data online, Videos S4, S5).

**Conclusion**

If the intra-operative transoesophageal echocardiography was not performed, our patient would have just had CABG, probably without solving the symptoms. In fact, no multiple fibroelastoma was detected by transthoracic echocardiography during her hospitalization. Furthermore, in future she would have undergone another cardiac operation for resection of aortic masses and valve replacement.

This incidental finding during intra-operative transoesophageal echocardiography may be useful to demonstrate how important is the role of these ultrasound images for the improvement of the diagnosis and the management in all cardiac surgical patients.

**Supplementary data**

Supplementary data are available at *European Journal of Echocardiography* online.

**Conflict of interest:** none declared.

**References**


Figure 3 High-power view of a transverse section of the papillary fibroelastoma at its origin from the valvular tissue. Some of the papillae show a circle of thin elastic fibres, coloured in purple with a Weigert-Trichromic stain (20×).