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CASE REPORT

Successful interdisciplinary treatment of a rare cause of acute myocardial ischaemia from intermittent tumour-associated obstruction of the left main coronary artery: a case report

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Background A papillary fibroelastoma of the aortic valve has been reported as a rare cause of myocardial ischaemia. An advanced combined interventional and surgical approach leading to sufficient therapy for the patient is presented in this case report.

Case summary A 56-year-old female patient presented in an emergency room of a hospital with an acute coronary syndrome. Over 1.5 years, recurrent stable angina had been known in the patient and significant coronary artery disease has already been ruled out in a previous coronary angiogram. The patient was immediately transferred to the catheter laboratory due to cardiogenic shock where a drug-eluting stent was implanted to, firstly, recanalize the left main coronary artery (LMCA) and, secondly, to protect the left main ostium from obstruction by an echocardiographic-proven mass. During subsequent deterioration of haemodynamics caused by decreasing left ventricular function and acute severe mitral insufficiency, firstly an intra-aortic balloon pump and secondly a veno-arterial extracorporeal membrane oxygenation was established through the femoral vessels. The patient was transferred to our cardiac surgery unit and was successfully operated utilizing a valve-sparing technique by extracting the tumour mass from the left coronary cusp and extracting the stent carefully from the LMCA. Histology revealed a papillary fibroelastoma.

Conclusion A papillary fibroelastoma of the aortic valve with intermittent obstruction of the coronary arteries requires surgical therapy. Interventional recanalization and extracorporeal support might be useful strategies to ensure the patient's safety as a bridge to surgery.

Keywords Heart valve tumour • Myocardial infarction • ECMO • Papillary fibroelastoma • Case report

Learning points

- Papillary fibroelastoma may be a rare cause for acute coronary syndrome.
- A combined interventional therapy and mechanical circulatory support may bridge haemodynamic instable patients for surgery.
- Undulating idiopathic anginous symptoms should lead to extended cardiologic diagnostic.

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Introduction

Cardiac tumours leading to ischaemia are one rare cause of myocardial ischaemia among various others. Heart valve associated tumours such as the benign papillary fibroelastoma might cause valvular defects, cardio-embolic events such as cerebral infarction or myocardial ischaemia due to coronary obstruction.[1,2]

In this report, we present the case of a papillary fibroelastoma-associated obstruction of the left coronary artery, initial cardiogenic shock, and the following treatment process.

Timeline

>1.5 years before	Anginal symptoms, intermittent and infrequent cardiological examination: electrocardiogram (ECG), transthoracic echocardiography (TTE) unremarkable
11 months before	Progressive anginal symptoms Unremarkable coronary angiography, ECG, and TTE
Day 0	Stress ECG with angina and ST-elevation Coronary angiography: tumour mass with intermittent obstruction of the left main, transoesophageal echocardiography: mobile mass, cardiogenic shock, intubation, intra-aortic balloon pump, interventional recanalization (two drug-eluting stent in left main), veno-arterial extracorporeal membrane oxygenation (ECMO) implantation, inter-hospital transportation, cardiac surgery (mass extraction)
Day 2	ECMO explantation
Day 5	Extubation
Day 14	Discharge

Case presentation

A 56-year-old female patient was admitted to an emergency room of an external hospital with symptoms of an acute coronary syndrome.

Approximately 11 months before, the patient had undergone coronary angiography ruling out obstructive coronary artery disease. Prior to this event the patient suffered from stress dependent but stable angina pectoris. According to the available medical data, both echocardiography and electrocardiography were reported as unremarkable. Despite a conservative therapeutic attempt with beta-blockers, statins, and anti-anginal ranolazine the patient suffered of combined physical and psychical symptoms and cardiac rehabilitation was started. Stenocardia and temporary ST-elevation in I, aVL, V1, V2 occurred during the routinely performed stress electrocardiogram at 125 W (*Figure 1A*). The patient was immediately admitted to the nearest cardiology hospital for further investigations. With suspected acute coronary syndrome, coronary angiography via a transradial approach was directly performed.

Coronary angiography revealed non-significant obstructed coronaries, not explaining her symptoms, especially since there was no main coronary stenosis. However, during the procedure either thrombus-like or tumour-like mass was seen, which seemed to occlude the left main coronary artery (LMCA) intermittently (*Video 1*). Simultaneous invasive arterial pressure monitoring revealed temporary relevant negative impact of her circulatory situation concurrent with the instable mass.

The platelet aggregation inhibitors tirofiban and acetylsalicylic acid were started, after percutaneous transluminal coronary angioplasty (balloon 2.0 mm × 12 mm), the thrombus mass was further detectable. Due to progressive haemodynamic instability, a drug-eluting stent (DES) (Promus 3.5 mm × 8 mm) was implanted, with the intention to re-open the LMCA (*Figure 1B and C* and *Videos 2 and 3*). This manoeuver was unfortunately not successful. The patient required immediate intubation, mechanical ventilation, and catecholaminergic support.

Our tertiary heart centre was contacted. Due to haemodynamic instability, conventional transportation to our hospital (>45 km) was not an option. Our mobile extracorporeal membrane oxygenation (ECMO) team was activated and immediately set out for the external hospital.

A transoesophageal echocardiography verified a mass originating from the left coronary cusp (LCC) and with a more or less cardiac-cycle synchronic prolapse into the direction of the left coronary ostium (*Figure 2* and Supplementary material online, *Videos S4 and S5*). The left ventricular (LV) function was highly impaired, with severe functional mitral insufficiency, and therefore an intra-aortic balloon pump (IABP) was implanted via the left femoral artery. An additional attempt to keep the mass away from the LMCA was tried utilizing an additional DES (Promus 4.0 mm × 16 mm) from the left coronary ostium in the aorta reaching just under the margin of the mass. With this manoeuver the blood flow in the LMCA was temporary ensured (*Figure 1D and E*) and further deterioration of the persistent unstable haemodynamic situation was avoided.

Approximately 60 min after first contact, our ECMO team arrived at the hospital. The IABP was discontinued and replaced by a peripheral veno-arterial (VA)-ECMO cannulation via the left femoral artery and right femoral vein including an additional left arterial distal leg perfusion. Veno-arterial ECMO support was started leading to notably improved haemodynamics. The patient was then transported to our hospital under stabilized conditions.

After arrival the patient was directly transferred into the operating theatre (*Figure 3*). After median sternotomy, cardioplegia infusion, and aortic clamping were applied, aortotomy was performed. Inspection of the aortic valve revealed a solid mass originating from the LCC. Measuring ∼2 cm × 1 cm in diameter and its main body localized close to the left main coronary ostium (*Figure 3B*). At the LMCA ostium the interventional implanted stent bodies were protruding from intraluminal into the aorta with the furthest end above the mass. After careful inspection, the mass was carefully retrieved (*Figure 3C*). After this, the native aortic valve was unremarkable and

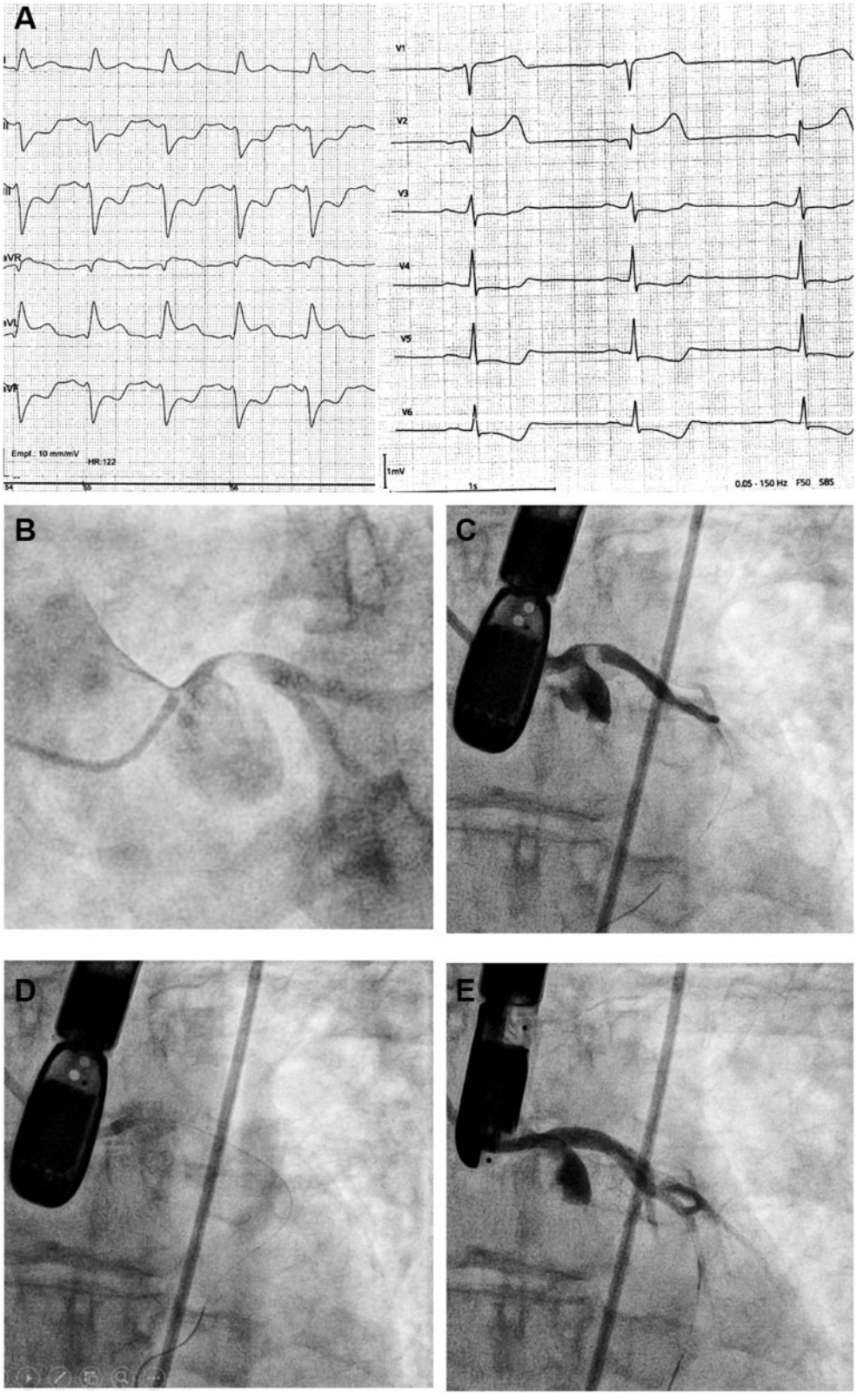
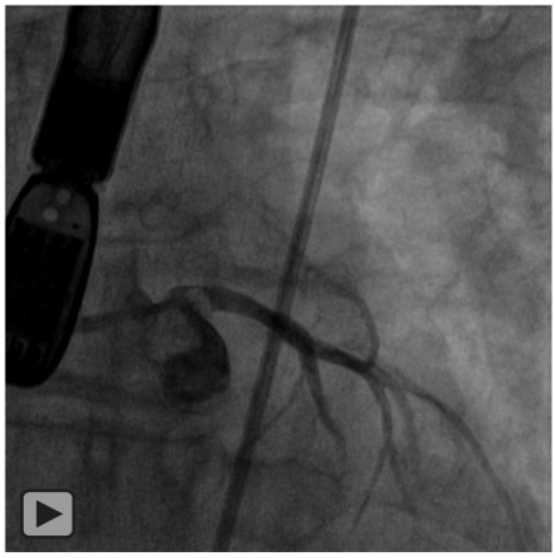
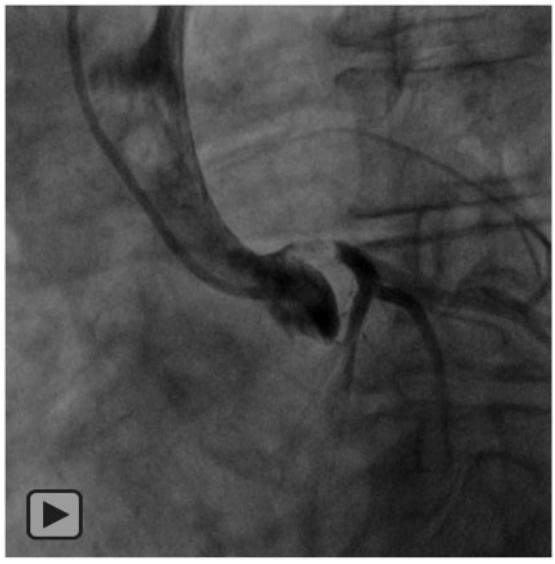


Figure 1 (*A*) Stress electrocardiogram with ST-elevation, (*B*) left main stem with contrast notch caused by the tumour mass, (*C*) left main coronary artery after initial drug-eluting stent implantation, (*D*) left main coronary artery with two drug-eluting stent, and (*E*) left main coronary artery after two drug-eluting stent implantation with momentary re-opened coronary.

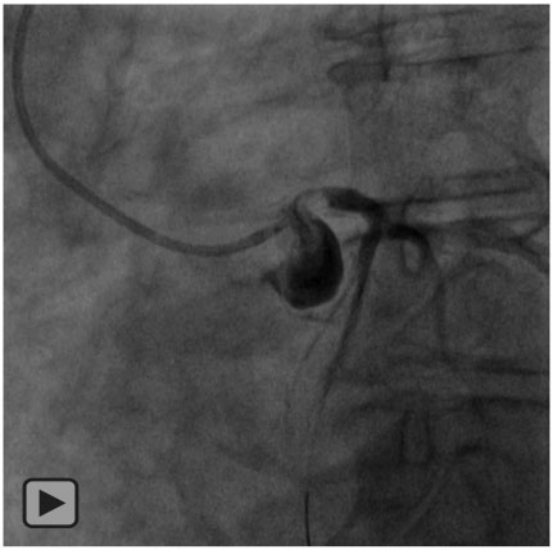


Video 1 Left main coronary artery obstruction by the tumour mass.



Video 2 Left main coronary artery with guiding wire, pre-drug-eluting stent implantation.

there was no indication for further surgical procedures. The implanted LMCA stents could be carefully retrieved without relevant visible damage such as dissection (*Figure 3D*). Echocardiography revealed a normal aortic valve function, without signs of insufficiency or valve damage. Since the patient required a higher dose of catecholamine and inotropic support, the heart–lung machine was switched again to VA-ECMO at the end of the operation. Under continuous extracorporeal support, sedation, and invasive ventilation, the patient was postoperatively treated in the intensive care unit. In the following hours the haemodynamic situation improved and slight weaning of VA-ECMO was started. There was no relevant blood loss associated with the cardiac surgery, acetylsalicylic acid therapy was continued (100 mg daily for 3 months). During the first postoperative day (POD), a transfusion-relevant GI-bleeding occurred, the small gastric bleeding spot was successfully treated by interventional coagulation therapy. The ECMO cannulas were successfully removed at the second POD. Weaning from the ventilator was complicated by purulent pneumonia. Extubation was possible on Day 5. Incipient symptoms of postoperative delirium were treated with dexmedetomidine. Fortunately, persistent neurologic symptoms were not detectable. Furthermore, in the postoperative course the patient did not suffer from any new anginal symptoms. Postoperative transthoracic echocardiography showed a competent aortic valve and an improved LV function (LV ejection fraction 58%, absence of regional wall movement disorder, MI I°, and compensated right ventricular function). The histopathologic examination revealed the mass as papillary fibroelastoma (*Figure 4*). The patient was discharged at the 14th POD to a rehabilitation centre.



Video 3 Left main coronary artery with guiding wire.

Discussion

A papillary fibroelastoma is a rare cause for acute myocardial ischaemia as it has been previously reported.[1–6] However, we believe that several features of our case are worth reporting: at least two aspects make this case special. First, the timing of the symptoms and the diagnosis. The patient suffered from at least over 1.5 years of anginal symptoms and was examined in two different cardiology departments. The cause of the symptoms was at that time not detectable. Retrospectively, it is not possibly to clearly differentiate whether the tumour mass was at that time too small to identify or if it was overseen. In summary, it is fair to say that the recurrent anginal symptoms led to a clear psychosomatic burden in our otherwise cardiac healthy patient.

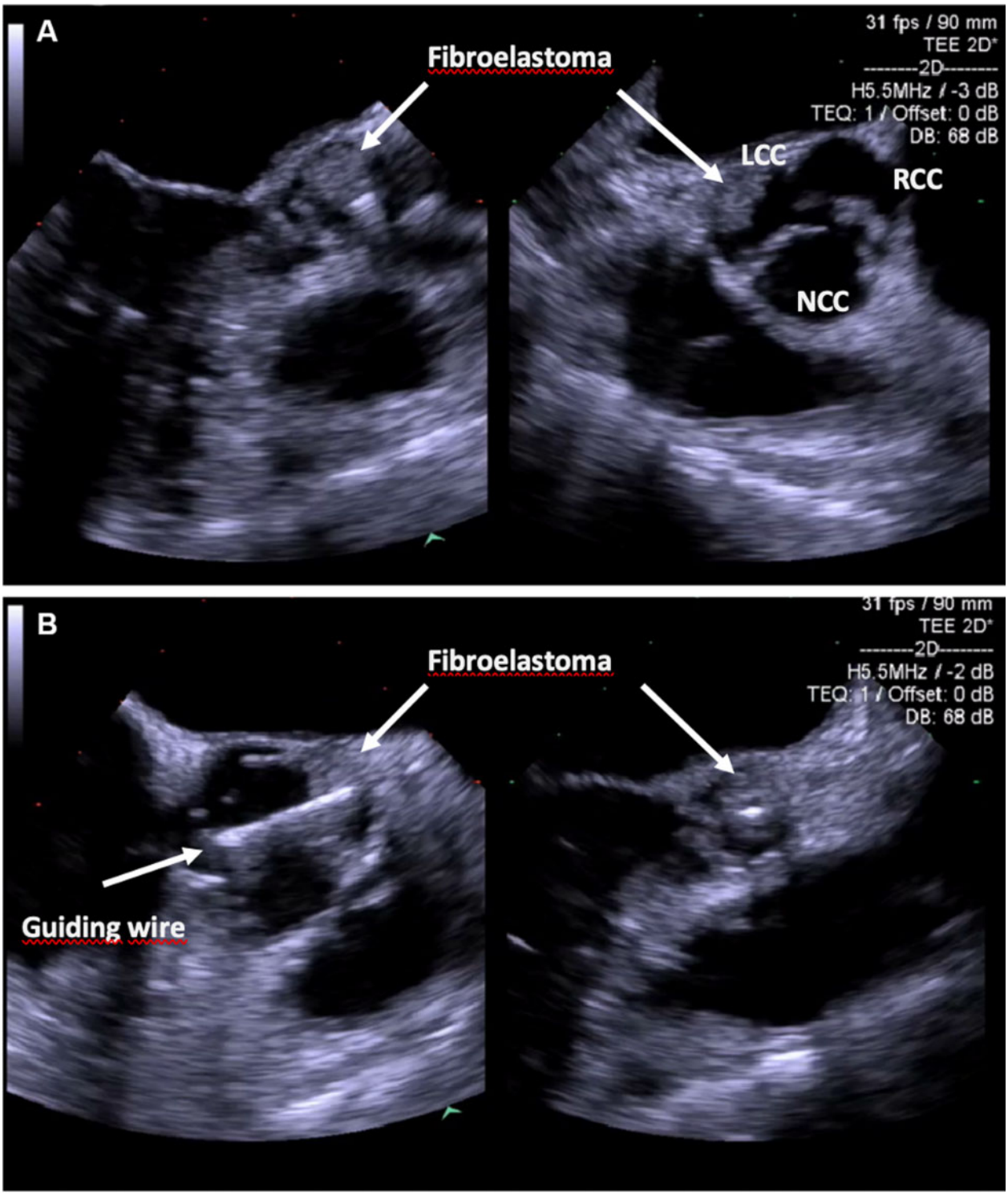


Figure 2 (*A*) Suspected tumour mass at the aortic valve and (*B*) transoesophageal echocardiography during left main coronary artery intervention with coronary guiding wire.

Second, the handling of the patient's situation. The interventional cardiologist's choice to try to keep the LMCA open with stent implantation allowed the patient to haemodynamically stabilize until definitive surgical therapy was possible. In order to minimize the mechanical stress on the implanted stent material, a second stent was implanted. The stent manoeuver is comparable to the chimney

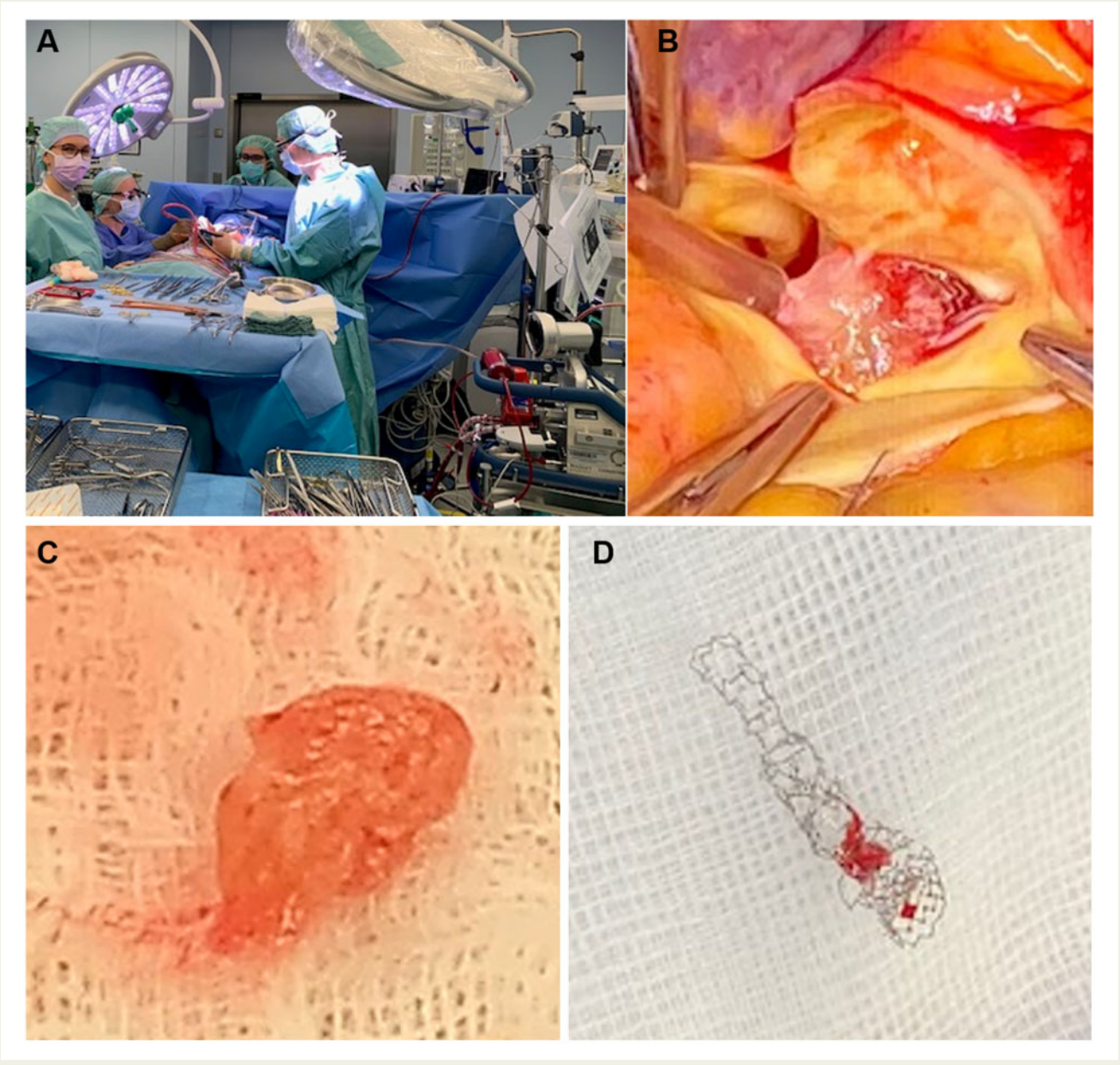


Figure 3 (*A*) intraoperative setting with ongoing veno-arterial extracorporeal membrane oxygenation support, (*B*) papillary fibroelastoma originating at the aortic valve, (*C*) excised tumour, and (*D*) explanted two drug-eluting stent of the left main coronary artery.

snorkel manoeuver which is sometimes performed in coronary obstruction during transcatheter aortic valve implantation (TAVI) procedures. The bridge-to-surgery was additionally complemented with VA-ECMO support allowing safe transfer to cardiac surgery.

Cardiac tumours are not very frequent. But especially in case of the rare papillary fibroelastoma (5% of all cardiac tumours[1,2]) treatment strategies are controversially discussed. If the benign tumour is found incidentally without clinical symptoms and it is small, a sometimes applied strategy is conservative observation. However, if the tumour mass is >1 cm, an increased risk for stroke through embolization or myocardial infarction with possible sudden cardiac death is described and a primary surgical therapy should be performed independent of the patient's symptoms.[7,8] In most cases a valve-sparing technique can be conducted. Recurrence of a once removed papillary fibroelastoma cannot be excluded, a lifelong cardiological checkup with routinely performed echocardiography is therefore suggested.

In patients with unexplained angina pectoris further and precise diagnostics are required, sometimes the rare cause could be a cardiac tumour, such as a papillary fibroelastoma. The interventional approach to keep the tumour mass away from the coronary ostia and the using extracorporeal support as bridge to surgery might be useful tools to ensure the maximum safety for those patients suffering from acute myocardial ischaemia.

local presentation is available online as Supplementary data

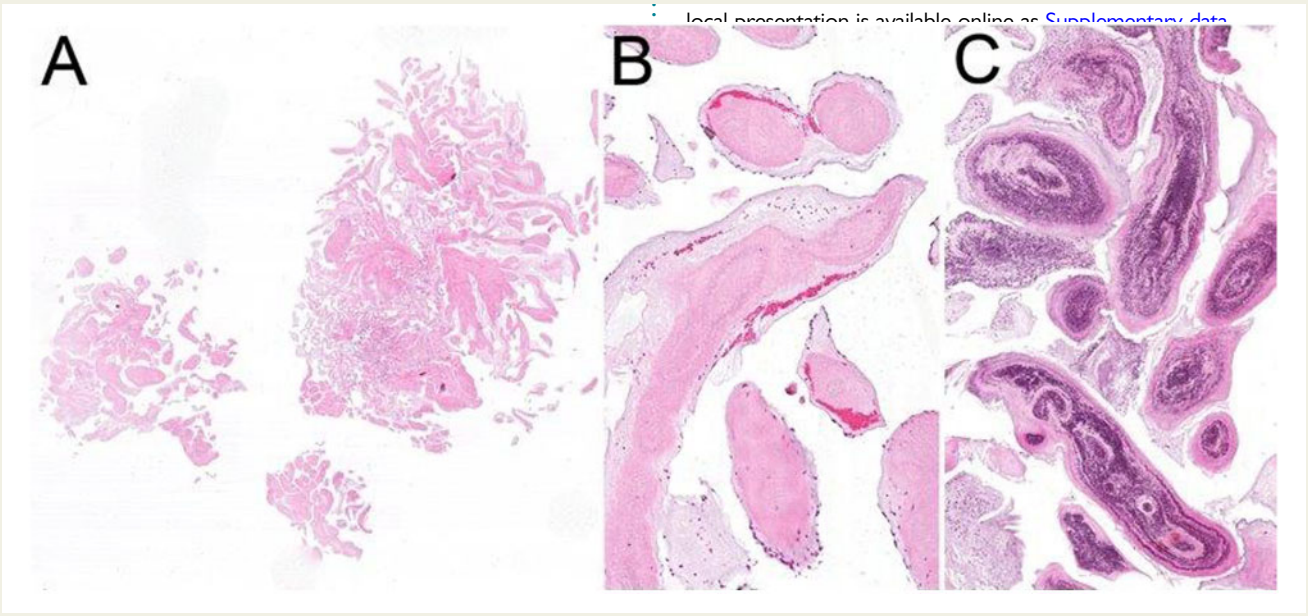


Figure 4 Histopathological examination of the papillary fibroelastoma with a fibroelastic core covered by a flat endothelium. Presence of flat hyalinoses. Absence of inflammatory or atypical signs. (*A*) Haematoxylin–eosin; 6.9-times magnification. (*B*) Haematoxylin–eosin; 100-times magnification. (*C*) Gieson's stain; 100-times magnification.

Lead author biography

Dr Katharina Huenges started her training as a cardiac surgeon in 2014 after graduating from Kiel University. The main interest of research and clinical focus is heart failure, transplantation, and extracorporeal support as well as transcatheter-based therapies.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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