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Intravascular Ultrasound-Guided Stent Implantation in the Left Main Coronary Artery Extrinsic Compression by Pulmonary Artery Aneurysm Due to Eisenmenger Syndrome

Eisenmenger Sendromunda Pulmoner Arter Anevrizmasına Bağlı Sol Ana Koroner Arter Basısına Intravasküler Ultrason Eşliğinde Stent Implantasyonu

ABSTRACT

Pulmonary arterial hypertension (PAH) is a profoundly destructive condition marked by the gradual narrowing and restructuring of small pulmonary arteries, leading to a rise in pulmonary vascular resistance (PVR), causing right-sided heart failure and, ultimately, mortality. During more advanced stages of this disease, patients may present with rare manifestations of pulmonary artery aneurysm (PAA) which are exertional chest pain, and hoarseness. The left main coronary artery compression (LMCA-Co) has been an increasingly recognized and possibly life-threatening entity in patients with severe PAH. The lack of well-established decision-making strategies for the management of both PAA and LMCA-Co has been remained as an unsolved issue in this setting. In this report, we present a case of percutaneous intervention of LMCA-Co with the guidance of intracoronary imaging in a patient with patent ductus arteriosus-Eisenmenger syndrome. Percutaneous intervention with intravascular guidance appears to be a safe and effective option for relieving symptoms and achieving positive clinical outcomes in patients with LMCA-Co.

Keywords: Left main coronary artery, percutaneous coronary intervention, pulmonary artery aneurysm, pulmonary hypertension

ÖZET

Pulmoner arteriyel hipertansiyon (PAH) küçük pulmoner arterlerin daralması ve yeniden yapılanması ile karakterize edilen, pulmoner vasküler direncin (PVD) artmasına, sağ kalp yetersizliğine ve en sonunda ölüme neden olan son derece yıkıcı bir hastalıktır. Bu hastalığın ileri evrelerinde hastalar, pulmoner arter anevrizmasının (PAA) nadir belirtileri olan efor ilişkili göğüs ağrısı ve ses kısıklığı ile başvurabilirler. Ağır PAH olgularında sol ana koroner arter basısı, giderek daha fazla tanınan ve yaşamı tehdit eden bir durumdur. Hem sol ana koroner arter basısı hem de PAA olan hastaların yönetim stratejisinde eksiklikler, hala çözülmemiş bir sorundur. Bu bildiride, patent duktus arteriosus-Eisenmenger sendromlu bir hastada intrakoroner görüntüleme rehberliğinde sol ana koroner arter basısının perkütan girişimini sunmaktayız. İntravasküler görüntüleme rehberliğinde perkütan girişim, sol ana koroner arter ekstrinsik basısı olan hastalarda semptomları hafifletmek ve olumlu klinik sonuçlara ulaşmak için güvenli ve etkili bir seçenek gibi görünmektedir.

Anahtar Kelimeler: Sol ana koroner arter, perkütan koroner girişim, pulmoner arter anevrizması, pulmoner hipertansiyon

CASE REPORT
OLGU SUNUMU

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Pulmonary arterial hypertension (PAH) is a profoundly destructive condition marked by the gradual narrowing and restructuring of small pulmonary arteries, leading to a rise in pulmonary vascular resistance (PVR), causing right-sided heart failure and, ultimately, mortality. The cardinal symptoms of PAH are dyspnea on exertion, fatigue, palpitations, edema, and syncope. During more advanced stages of this disease, patients may present with rare manifestations of pulmonary artery aneurysm (PAA) which are exertional chest pain, and hoarseness.[1] Identifying the symptoms of extrinsic compression of the left main coronary artery compression (LMCA-Co) by PAA in patients with PAH can be challenging due to their rarity and resemblance to other

cardiac symptoms. More than 50% of the cases of PAA have been associated with congenital heart diseases and pulmonary hypertension, particularly patent ductus arteriosus (PDA).[2]

The lack of well-established decision-making strategies for the management of both PAA and LMCA-Co has been remained as an unsolved issue in this setting. In this report, we present a case of percutaneous intervention of LMCA-Co with the guidance of intracoronary imaging in a patient with PDA-Eisenmenger syndrome, along with a concise review of the literature.

Case Report

A 38-year-old female diagnosed with PDA and Eisenmenger syndrome 9 years earlier was referred to our clinic with syncope, progression of dyspnea, and new-onset chest pain on exertion. Her saturation was 87% on room air, electrocardiogram showed sinus rhythm with right axis deviation, the right ventricular strain pattern, and incomplete the right bundle branch block (Supplementary Figure 1). Her 6-min walking distance was 260 meters and pro-BNP level was 2341 ng/L. She was on triple therapy with macitentan, tadalafil, and selexipag. Imaging studies including transthoracic echocardiography (TTE) and computed tomography angiography confirmed PDA with bidirectional shunt and PAA of 4.7 cm, markedly enlarged right heart chambers, flattening of the interventricular septum, moderate tricuspid and pulmonary regurgitation, and preserved right and left ventricular functions (Supplementary Figure 2). Due to debilitating anginal symptoms and high clinical suspicion of LMCA-Co, the patient was taken to a catheterization laboratory for invasive imaging. Coronary angiogram (CAG) of left anterior oblique (LAO) 45° and LAO cranial 30° views demonstrated severe occlusion of the proximal LMCA and the LMCA take-off angle in reference to the left sinus of Valsalva was <30° (Figure 1A-C). Intravascular ultrasound imaging was performed to evaluate the LMCA and resulted in slit-like appearance of the LMCA and severe dynamic obstruction, minimal lumen area (MLA) of 3.6 mm^2 without any atherosclerotic component (Figure 1D). Therefore, percutaneous coronary intervention (PCI) of the LMCA was decided by the local heart team. The LMCA was engaged with a Judkins guiding catheter, both the left anterior descending artery and the circumflex artery were wired using a 0.014'' floppy guidewires. A 4.5 × 9 mm bare metal stent was directly implanted in the proximal LMCA (Figure 2A). Post-PCI IVUS imaging revealed partial underexpansion of the stent and consequently post-dilated with a 4.5 mm non-compliant balloon. The final IVUS assessment demonstrated full expansion of the stent and adequate apposition to the vessel wall (Figure 2A and B). The patient was discharged on the following day with low-dose aspirin and clopidogrel in addition to triple PAH-specific treatment. During the 1st-month follow-up, the patient remained on the World Health Organization functional Class II and did not report chest pain anymore.

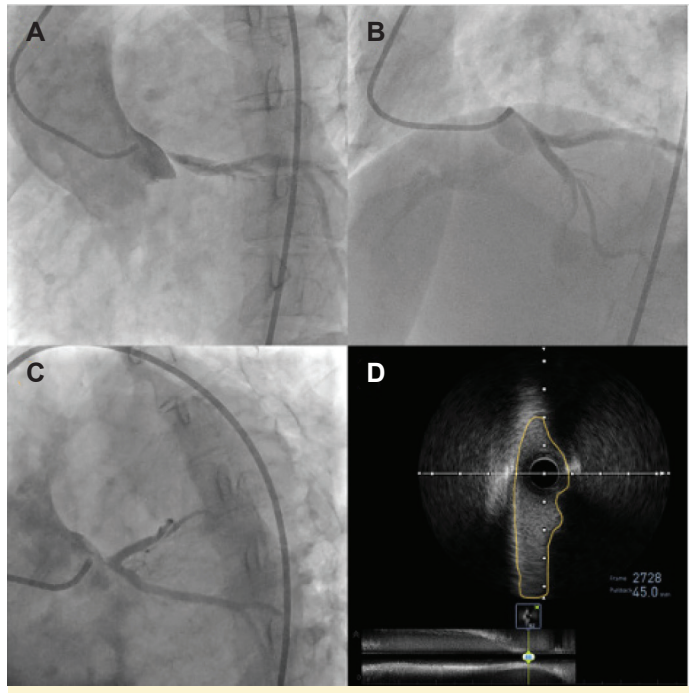

Figure 1. (A) Coronary angiogram in the left anterior oblique (LAO) and (B) LAO cranial views demonstrated subtotal occlusion of the proximal left main coronary artery (LMCA) and the LMCA take-off angle in reference to the left sinus of Valsalva was <30°. (C) LAO caudal (spider) view might be misleading due to the ovoid-shaped deformation of the LMCA. (D) Intravascular ultrasound imaging demonstrating slit-like appearance of the LMCA and severe dynamic obstruction, minimal lumen area of 3.6mm² without any atherosclerotic component.

Discussion

Since the initial description of LMCA-Co as a rare complication nearly 70 years ago, there has been a growing focus on the treatment and management of this potentially lethal condition.[3] This condition holds considerable prognostic significance and can lead to myocardial ischemia, left ventricular dysfunction, arrhythmias, and sudden death.[3] Typically defined as >30 mm in size, PAAs are primarily associated with pulmonary hypertension in the background of congenital heart diseases. The most common underlying conditions include PDA, ventricular septal defect, and atrial septal defect.[2]

The first line non-invasive imaging tool remains TTE, offering valuable assistance in the measurement of PAA diameter and determining the probability of LMCA-Co. It has been reported that LMCA-Co was not seen in patients with PAA <40 mm; moreover, different cutoffs for the PAA to aortic diameter ratio, ranging from 1.2 to 2, have been proposed for evaluation in this context.[3-6] In the largest prospective single-center series, PAA diameter larger than 40 mm was found to be the strongest predictor for LMCA-Co.[6] Thus, a stepwise approach starting with TTE and moving further with multidetector computed

ABBREVIATIONS

CAG	Coronary angiogram
LAO	Left anterior oblique
LMCA-Co	Left main coronary artery compression
MLA	Minimal lumen area
PAA	Pulmonary artery aneurysm
PAH	Pulmonary arterial hypertension
PCI	Percutaneous coronary intervention
PDA	Patent ductus arteriosus
PVR	Pulmonary vascular resistance
TTE	Transthoracic echocardiography

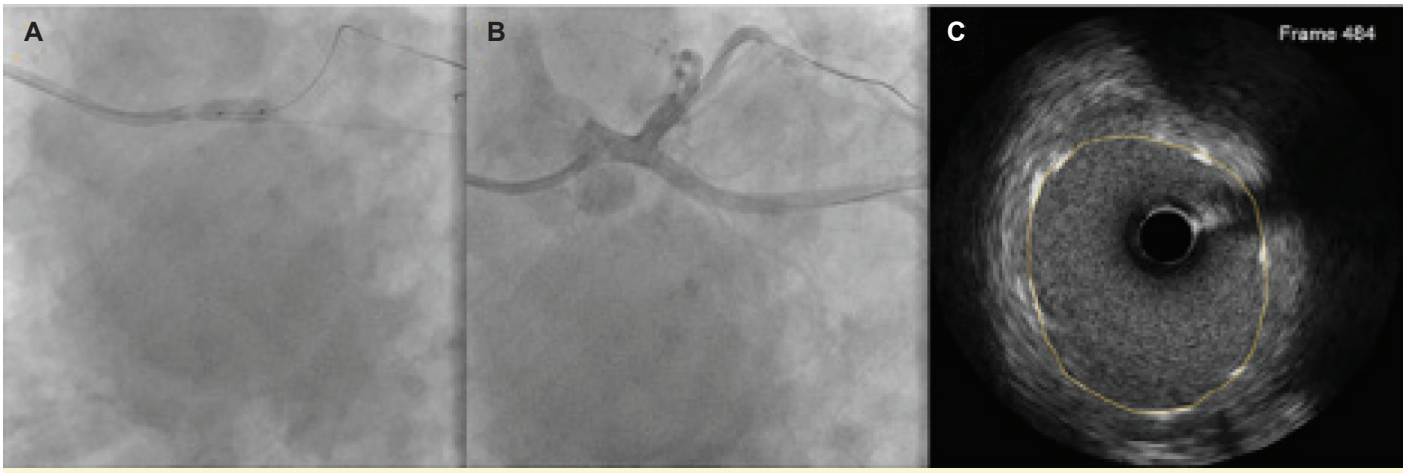

Figure 2. (A) 4.5×9 mm bare metal stent was directly implanted in the proximal LMCA. Final angiogram (B) and IVUS imaging (C) demonstrated full apposition and expansion of the stent.

tomography (MDCT) or CAG could be a feasible strategy. In the same study comprising 765 patients, it was reported that approximately 16% of patients underwent MDCT due to angina or angina-like symptoms. Cardiac MDCT offers a comprehensive approach to assess various aspects, including the severity of extrinsic LMCA-Co (>50%), the angulation between the LMCA and the left sinus of Valsalva, the underlying pathology of PAA, as well as left and right ventricular functions. Within the overall patient cohort, LMCA-Co was angiographically confirmed in 6% of cases.[6] In previously reported series of Akbal et al. [5], LMCA-Co >50% was noted in 8.2% of patients who underwent CAG, and in 4.4% of overall PH patients, emphasizing the significant prevalence of this potentially life-threatening complication. In addition, not only PA diameter and PA diameter to aortic diameter ratio but also the clinical etiology of PDA, a younger age, and severity of PAH as defined by higher PA mean pressure and PVR predicted the risk of LMCA-Co.

In patients with high clinical probability of LMCA-Co and presenting supportive non-invasive evidence, CAG should be pursued. LMCA-Co is best visualized in the LAO and LAO with cranial angulations and can be differentiated from atherosclerotic plaque with downward angulated take-off of LMCA ostium and eccentric narrowing progressing to a smooth normalization in the distal region. IVUS can offer additional information on the presence of atherosclerosis, exposing dynamic compression of the LMCA by PAA, optimizing stent placement and adequate stent expansion during intervention.[3] Velázquez Martín et al. [7] concluded that IVUS allows better discrimination of significant extrinsic LMCA compression due to a PAA in PH patients when compared with CAG, avoiding unnecessary treatment in 50% of cases with positive angiographic criteria for LMCA-Co (>50%). Moreover, generally accepted MLA cutoff of >6 mm^2 can be safely adopted as a criterion to postpone revascularization in PAH patients with LMCA-Co.[7]

Although not established due to the limited number of cases, LMCA stenting has emerged as the preferred revascularization strategy, demonstrating favorable angiographic outcomes and satisfactory short-term clinical results, and compression of the ostial or proximal part of this artery, sparing the LMCA bifurcation permits a single stent placement.[5,6,8,9] To date, the largest prospective series with the longest follow-up has been reported by Galiè et al.[6,8] Their results not only suggested high procedural success with a focus on safety but also reported low stent recoil, mechanical compression, and demonstrated favorable clinical outcome at long-term follow-up. During 5-year follow-up, only five of 53 patients had in-stent restenosis and underwent PCI, and no cases of stent thrombosis were reported.[8] Correspondingly, Akbal et al.[5] demonstrated the safety of stenting with positive clinical outcomes in a series of 12 patients with a median follow-up of 17.3 months. There was only one non-sudden cardiac death reported on 10th day after stenting, and no mortality during follow-up.[5] In series of Velázquez Martín et al. [7], there was no sudden death, in-stent restenosis or bleeding complication related to antiplatelet/anticoagulant treatment among eight patients who underwent stenting. Moreover, recent meta-analysis of Badea et al. [9] including five small-sized cohorts and 64 case reports revealed significant anginal symptom relief and a low mid-term mortality rate associated with coronary stenting. Stent choice varies across different series. In contrast to the 48.9% rate of drug-eluting stent implantation in the series of Galiè et al.,[6,8] Akbal et al. [5] favored bare metal stents in all patients. On the other hand, all of the patients underwent drug-eluting stent implantation in the series of Velázquez Martín.[7] Beside these studies, the majority of reported cases have consistently favored the use of drug-eluting stent as the stent choice.[9,10] In addition, ensuring long-term radial force resistance against external compression presents a noteworthy challenge in this context. The benefit of using drug-eluting stent in this setting has yet to be established due to the non-atherosclerotic obstruction of the LMCA. Given the similar rates of in-stent restenosis, stent thrombosis, and sudden death observed among patients implanted with drug-eluting stent or bare metal stent, stent choice might be left to operator's discretion, taking into account factors such as the patient's need for additional anticoagulation treatment or the presence of a high bleeding risk.

Surgical approach might be considered in selected patients, particularly in those requiring surgery for another concurrent

condition. Unlike aortic aneurysms, there is no clear consensus on management strategy and surgical indications for PAAs. In adults, surgical repair of PAA has been considered for individuals with the following indications: Main PA diameter >5.5 cm, an increase in PAA diameter of more than 5 mm within 6 months, presence of clinical symptoms, severe coexisting valvular pathologies or shunt flow, verification of LMCA-Co or adjacent structures, and thrombus formation in the PA.[2] In the aforementioned cases, aneurysmorrhaphy or heart-lung transplantation remains the current choice of treatment.[2]

Conclusion

LMCA-Co has been an increasingly recognized and possibly life-threatening entity in patients with severe PAH. Percutaneous intervention with intravascular imaging guidance appears to be a safe and effective option for relieving symptoms and achieving positive clinical outcomes in patients with LMCA-Co.

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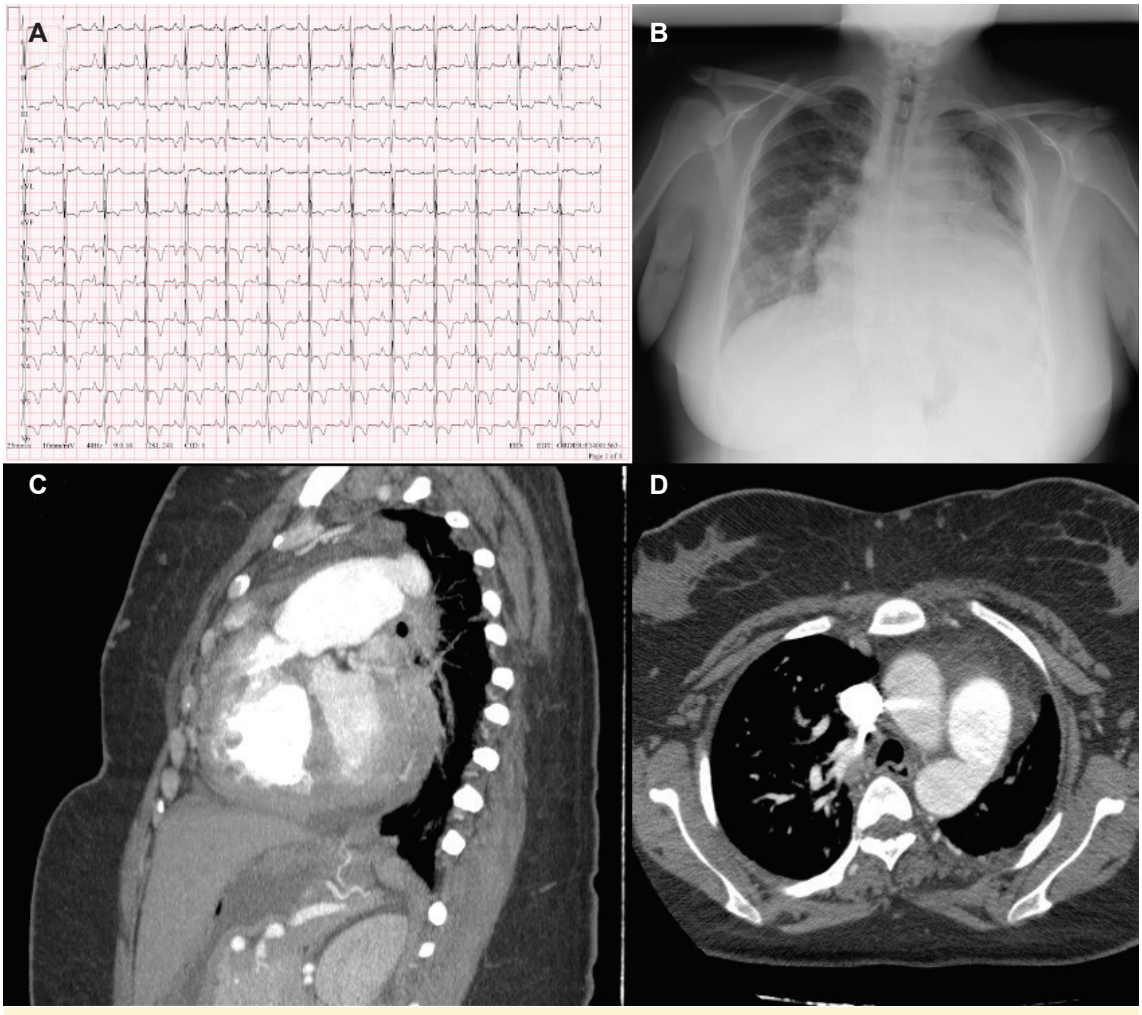

Supplementary Figure 1. (A) Electrocardiogram demonstrating sinus rhythm with right axis deviation, right ventricular strain pattern, and incomplete right bundle branch block. (B) Chest X-ray showing enlarged right heart chambers and pulmonary conus. (C) Sagittal and (D) axial views of computed tomography angiography showing patent ductus arteriosus.

Supplementary Figure 2. Transthoracic echocardiogram demonstrating (A) pulmonary artery aneurysm of 4.7 cm, (B) flattening of the interventricular septum, and (C) estimated pulmonary artery pressure of 87 mm Hg.