Type 1 neurofibromatosis (NF1), also known as von Recklinghausen disease, is an autosomal dominant multisystem genetic disorder. Vascular manifestations of NF1 in coronary arteries include aneurysmal and stenotic lesions, which may predispose to thrombotic mass formation, and eventually, myocardial infarction (MI). The appearance of recurrent stent thrombosis in patients with NF1 has not been reported so far.

A 55-year-old man was admitted to the emergency department due to severe chest pain lasting 4 hours. He was diagnosed with NF1 in childhood and had typical massive neurofibromas on the skin (FIGURE 1A) and a benign tumor in the brain (FIGURE 1B). He underwent elective percutaneous coronary intervention (PCI) of the left ascending artery in 2014 and the right coronary artery (RCA) in 2015 with drug-eluting stents implantation. In 2018, he was admitted with ST-segment elevation MI due to stent thrombosis within the RCA, which was successfully treated with manual aspiration thrombectomy followed by balloon angioplasty (FIGURE 1C). Dual antiplatelet therapy was modified to acetylsalicylic acid (75 mg once daily) and ticagrelor (90 mg twice daily). Ten months after stent thrombosis, he was admitted again due to inferior wall ST-segment elevation MI. Urgent coronary angiography revealed another stent thrombosis within the RCA (FIGURE 1D). PCI was performed with aspiration thrombectomy and glycoprotein IIb/IIIa inhibitor was administered intracoronary, followed by drug-eluting stent implantation (Xience PRO, 3.75 × 48 mm) at 18 atm. An optimal result was achieved with post-dilatation with noncompliant balloon, 3.75 × 20 mm, at 20 atm (FIGURE 1D). No complications were observed and left ventricular ejection fraction improved.
fraction at discharge was 41%. Dual antiplatelet therapy was sustained with acetylsalicylic acid and ticagrelor, as the neurologist denied the addition of oral anticoagulation due to brain tumor. Unfortunately, the patient suddenly died 1 year after the last PCI. Stent thrombosis could have been the cause of death, but neither angiography nor autopsy was performed.

So far, PCIs in patients with neurofibromatosis have been rarely reported. The presence of large coronary aneurysms is postulated as a reason for acute MI in this population. However, the pathophysiology of vascular pathologies in neurofibromatosis is multifactorial:3 from disturbed smooth muscle migration to impaired vascular tissue histogenesis and healing, which could be crucial after stent implantation and may explain repeated stent thrombosis despite optimal antiplatelet treatment in our patient.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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