Image in cardiology

An Unusual "Black" Transcatheter Aortic Valve Implantation

Implante percutáneo «negro» de válvula aórtica

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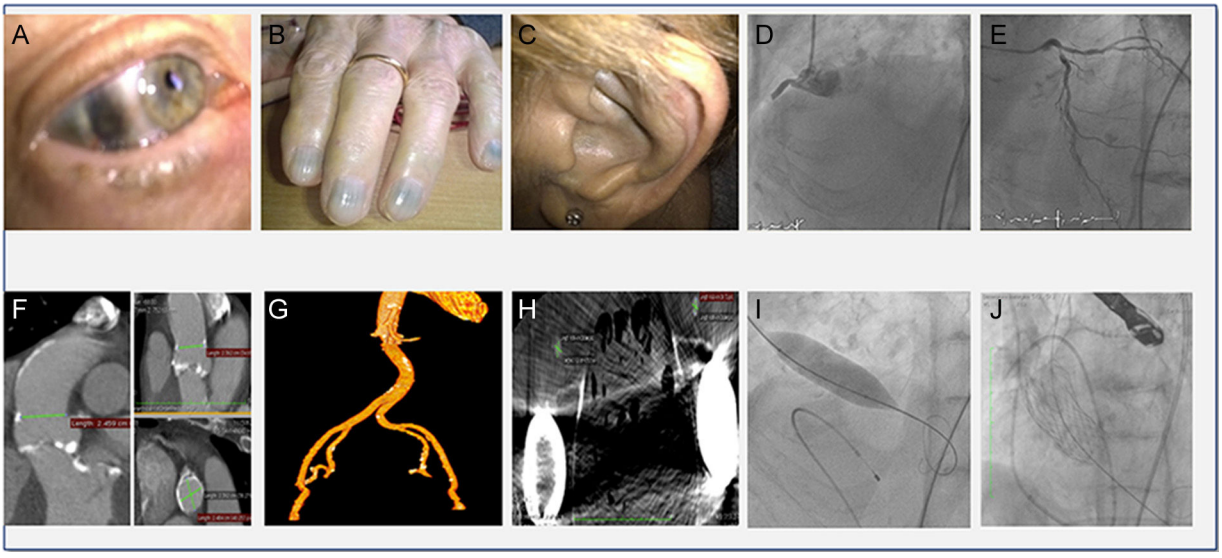

Figure.

A 78-year-old-woman was referred for inferior ST-segment elevation myocardial infarction. Physical examination revealed a 3/6 systolic ejection murmur and a bluish-black discoloration of the sclera, nails, and auricula (Figures A-C). Coronary angiography showed occlusion of the right coronary artery (Figure D) and critical stenosis of the left main and left anterior descending artery (Figure E). Primary right coronary artery angioplasty was performed. However, echocardiography revealed severe aortic stenosis and left ventricular dysfunction. The patient also had debilitating diffuse arthropathy with a previous bilateral hip prostheses. A diagnosis of alkaptonuria was established. Logistic EuroSCORE and Society of Thoracic Surgeons score were 45% and 25.44%, respectively. An intravascular ultrasound-guided unprotected left main-anterior descending artery drug-eluting stent was performed. A computed tomography scan was done before transcatheter aortic valve implantation (Figures F-H) and, due to hemodynamic instability, a bridge aortic balloon valvuloplasty was successfully carried out (Figure I and Video of the supplementary material) followed by implantation of a 29 mm self-expandable prosthesis (Figure J).

Alkaptonuria is an autosomal-recessive inherited disorder of tyrosine metabolism, with various systemic abnormalities related to ochronotic pigment deposition with collagen tissue degeneration and calcification. Cardiovascular manifestations of alkaptonuria often include aortic stenosis and rarely coronary artery disease. Cardiac surgery and anesthesia may be challenging due to hostile chest, porcelain aorta, and restrictive pulmonary disorder, with an increased risk of postoperative sternal dehiscence. In such high-risk frail patients, percutaneous coronary intervention and transcatheter aortic valve implantation may represent viable therapeutic options. However, the extensive aortic calcifications require accurate planning of the vascular access site with computed tomography angiography.

SUPPLEMENTARY DATA

Supplementary material associated with this article can be found in the online version available at https://doi.org/10.1016/j.rec.2017.10.006.

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