Aortic valve replacement surgery reveals previously undiagnosed alkaptonuric ochronosis

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A 64-year-old patient with aortic stenosis underwent surgical aortic valve replacement. At surgery, severe annular calcification and blackish-blue discoloration of the valve cusps, aortic wall and coronary arteries were noted (Fig. 1). Microscopic examination revealed dark ochronotic pigment in the valvular tissue (Fig. 2).

Figure 1: Ochronosis of the ascending aortic wall (A) and aortic valve annulus (B).

Figure 2: Ochronosis of the aortic valve leaflets (A) and HE staining of aortic valve leaflets (B and C) showing ochronotic pigment (arrows).